Guide to Pediatric Urology and Surgery in Clinical Practice

Prasad P. Godbole
Duncan T. Wilcox
Martin Koyle
Editors

Second Edition

Springer

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Introduction

It is with great pleasure that we introduce the second edition of the book ‘Guide to Pediatric Urology and Surgery in Clinical Practice’. This book is aimed at Primary Care Physicians, General Practitioners, Pediatricians, and trainees embarking on a career in pediatric urology or surgery.

With increasing demand and pressure on secondary and tertiary care services for pediatric urology and surgery, it is important that the resources available are put to best use. This book therefore highlights the most common pediatric urology and surgical conditions seen in primary care or by Pediatricians. Brief details about the condition, key points, and indications for referral to specialist Pediatric Urologists or Pediatric Surgeons and management in the nonspecialist setting are highlighted. This book does not replace exhaustive texts in Pediatric Urology or Pediatric Surgery but serves as a quick reference guide on managing these conditions in a nonspecialist setting.

We would like to thank the contributors for their timely submission of the chapters. We are grateful for the support of Melissa Morton, Executive Editor at Springer Nature and Vickrutha Sudarshan, Project Manager at SPi Global to enable us to stay on track in the publication of this book.

Finally it goes without saying that we are deeply indebted to our families who as always have been patient and supportive during the whole process.

Chapter 1
Urinary Tract Infection USA

Angela M. Arlen and Christopher S. Cooper

Key Points

- UTI must be suspected in febrile infants as symptoms may be nonspecific.
- Catheterized urine specimens are superior, but mid-stream clean catch samples can be obtained in toilet-trained children with proper instruction.
- Successful empirical treatment should be based on local epidemiology and susceptibility rates and adjusted according to organism and sensitivity profile.
- Neurogenic bladders managed with frequent instrumentation will have colonization and urine culture should be obtained only if the child is symptomatic.
- Toilet-trained children with UTI should be evaluated and treated for any bladder-bowel dysfunction.
- Evaluation of febrile UTI include renal-bladder ultrasound ± VCUG.

Introduction

Urinary tract infection (UTI) is a commonly diagnosed bacterial infection in childhood, accounting for nearly 1% of pediatric office visits and approximately 10% of emergency department visits. It is estimated that 7% of girls and 2% of boys under the age of 6 will be diagnosed with UTI, and infection may be the first

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symptom of a congenital anomaly of the urinary tract. Despite the potential for significant morbidity, diagnosis and management of pediatric urinary tract infection remains controversial. In addition to prompt diagnosis and appropriate antibiotic treatment, the goal of pediatric UTI management is to identify those children at risk for recurrent infection/parenchymal damage and to address any modifiable risk factors.

Risk Factors

There are a host of factors that make bacteriuria and subsequent sequela more or less likely for any given child.

- Age—prevalence of bacteriuria is highest in the first year of life
- Gender—first 12 months are the only time when males have more UTIs than females
- Race and ethnicity—Caucasians are more likely than Hispanic or African-American children to develop a UTI
- Circumcision status—circumcision reduces the UTI rate by tenfold in the first 6 months of life
- Urinary stasis from incomplete bladder emptying or infrequent micturition allows for growth of bacteria that would be normally be flushed out with voiding
- Dysfunctional voiding, i.e. “habitual contraction of the urethral sphincter during voiding” resulting in incomplete emptying
- Approximately one third of children with recurrent UTIs have associated stool retention/constipation with or without encopresis
- Genitourinary anomalies
    - Renal scarring
    - Vesicoureteral reflux
    - Hydronephrosis
    - Neurogenic bladder managed with intermittent catheterization—bacteriuria and pyuria are present in most children and *E. coli* strains often reflect colonization rather than symptomatic infection

Presentation

Infants and Young Children

- Clinical presentation tends to be nonspecific and reliable urine specimens for culture cannot be obtained without invasive measures (urethral catheterization or suprapubic aspiration)
- Symptoms correlate poorly with bacterial localization to upper urinary tract

- In a febrile infant (rectal temperature ≥38 °C), urine specimen should be obtained for urinalysis and culture prior to starting antimicrobial therapy
- UTI accounts for ~5 to 7% of fevers in this age cohort
- Other common symptoms include malodorous urine, irritability, lethargy, poor appetite, vomiting, and diarrhea

Older Children (>2 years of age)
- Common symptoms include dysuria, urgency, frequency, incontinence, malodorous urine, and suprapubic discomfort
- Other symptoms include nausea, vomiting, flank pain, and generalized malaise

Diagnosis

In infants and young children, a urine specimen must be obtained via urethral catheterization or suprapubic aspiration as the diagnosis of UTI cannot be reliably established with bag specimens. Bagged specimens are only useful if negative; if positive, a catheterized specimen must be obtained resulting in delay of diagnosis and treatment. Bagged specimens are unacceptable in high-risk patients including infants less than 2 months of age.

Urinalysis (UA) with microscopy
- Allows for presumptive UTI diagnosis but is not a substitute for culture
- Specimen must be fresh (<1 h at room temperature, <4 h refrigerated)
- Nitrite—highly specific when positive; limited utility in infants who empty bladder frequently as it takes time for bacteria to convert nitrate to nitrite
- Leukocyte esterase—highly sensitive but false positives are common
- Significant pyuria is defined as >5 WBC/hpf
- Bacteria visualized on fresh, uncentrifuged samples predictive of infection
- Positive leukocyte esterase or nitrite with pyuria is highly predictive of infection

Urine culture
- Greater than or equal to 50,000 CFU/mL with evidence of pyuria on urinalysis is the current standard for positive culture and UTI
- Clinicians should consider UTI in urine culture with >10,000 CFU/mL in catheterized specimens if fever and/or other symptoms or pyuria are present

Common Pathogens

The most common bacteria affecting the urinary tract are gram negative Enterobacteriaceae, most commonly *E. coli*. Bacterial surface structures such as pili or fimbriae increase virulence and uropathogenicity.

- *Escherichia coli* (54–67%)
- *Klebsiella* (6–17%)
- *Proteus* (5–12%)
- *Enterococcus* (3–9%)
- *Pseudomonas* (2–6%)

Treatment

When initiating antimicrobial therapy, route and antibiotic choice should be based on practical considerations. Goals of therapy include eradication of acute infection, prevention of complications, and minimize likelihood of renal damage and recurrent infections. Successful empirical treatment should be based on local epidemiology and susceptibility rates and adjusted according to the cultured organism and sensitivity profile. In children presenting with fever, 7–14 days of therapy is recommended.

Infants younger than 3 months of age
- Neonates require admission with full sepsis evaluation
- Need for admission with parenteral therapy is based on clinical status (toxic appearance, dehydration, electrolyte anomalies, PO intolerance)
- Neonates should be covered for *Enterococcus* species when choosing empiric antibiotics
- Febrile infection in this cohort progresses to urosepsis more frequently

Young Children (<24 months of age)
- Outpatient therapy with once daily intramuscular third generation cephalosporin
- If capable of taking oral fluids, 7 to 14 days of oral antibiotic with broad genitourinary pathogen coverage
- No follow-up culture is required if organism is sensitive to selected antimicrobial and symptoms resolve

Older Children
- If uncomplicated UTI, oral antibiotic agents are well-tolerated and clear the infection after a 3 to 5-day treatment course
- Children with fever should be treated for a minimum of 7 days

Imaging

All children with a culture-documented UTI and a fever of greater than 38° should undergo follow-up imaging to detect anatomic abnormalities. Various guidelines have been established for imaging the child with UTI, however all fall short of identifying only those patients who will benefit from the results of the imaging.

- Renal-bladder ultrasound should be obtained in all children with febrile UTI, males of any age with first UTI, and children with recurrent infections
- Voiding cystourethrogram
  - Anatomic imaging of bladder and urethra
  - Should be obtained in children with recurrent febrile infections and those with an abnormal screening ultrasound, after infection has cleared
  - Up to 40% of children with febrile UTI may have vesicoureteral reflux
- DMSA renal scintigraphy
  - Useful in high risk patients for prognostic purposes
  - ~15% of children with UTI have renal scarring on follow-up renal scan and the percentage increases as the number of UTIs increase

Indications for Referral

Children with recurrent febrile UTI should be referred to Pediatric Urology, as should those with a UTI and abnormal ultrasound or vesicoureteral reflux. Toilet-trained children with bladder and bowel dysfunction refractory to timed voiding and bowel regimen also warrant referral.

Suggested Reading

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Chapter 2
Abnormalities of the Inguinal Canal and Scrotum

Kunj R. Sheth and Paul F. Austin

Key Points

- The diagnosis of most acute abnormalities of the inguinal canal and scrotum can be made by the history and physical exam.
- Testicular torsion is a urologic emergency whereas torsion of the testicular appendages may be managed expectantly.
- Most other conditions causing an acute scrotum can be managed conservatively in primary care.
- A painless, non-acute testicular mass is considered a tumor until proven otherwise. The majority of pre-pubertal testicular tumors are benign.

Introduction

Abnormalities of the inguinal canal and scrotum are frequently encountered in children and adolescents. Certain conditions require emergent care while others are non-emergent. Sorting through the many different conditions that result in an "abnormal appearing" inguinal canal and/or scrotum is critical in determining the acuity of care and in choosing the appropriate treatment. This chapter will outline the different conditions that result in inguinal canal and scrotal abnormalities and assist the primary care provider in diagnosis and management.

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Common Abnormalities of the Inguinal Canal and Scrotum

1. Testicular torsion: Testicular torsion is a urologic emergency and typically occurs in pubescent boys but may also present in the pre-pubescent period. The diagnosis is primarily a clinical determination that is made from the medical history and the physical exam. Radiologic imaging with scrotal ultrasonography is beneficial when the diagnosis is in doubt. Key diagnostic symptoms and signs include: nausea, vomiting, acute onset, diffusely tender and palpably hard testis. Other helpful findings include a high-riding testis (Fig. 2.1), a horizontal lie of the testis and no cremasteric reflex. A colicky type of pain is typically seen early in the time course of testicular torsion. With delayed presentation, no tenderness in a palpably hard testis is an ominous sign. Torsion is most commonly seen in the peri-pubertal population, but can present in prepubertal patients, often with a more difficult diagnosis.

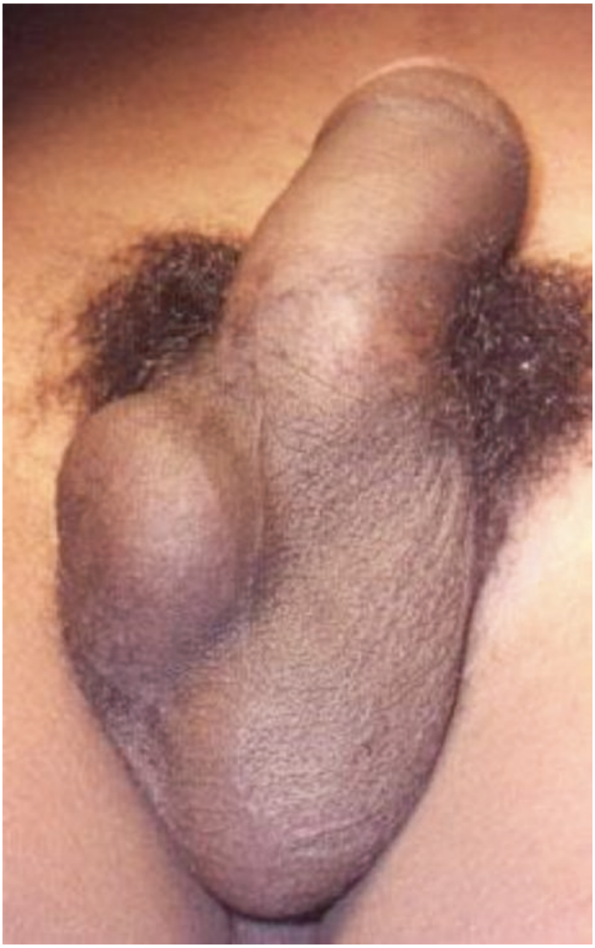

Fig. 2.1 High riding right testis

2. Torsion of the appendix testis or epididymis: Torsion of the appendage of the testis or epididymis is common in the prepubescent child and is a great "mimicker" of testicular torsion. The onset of pain is usually acute, but may develop slowly over time. The intensity of pain ranges from mild to severe. The pain is generally located in a specific location such as at the superior pole of the testicle corresponding to the most common location of the appendix testes. However, with prolonged duration, diffuse tenderness develops from the resultant inflammatory response. The pain is worse with movement & better with rest. If left untreated, diffuse scrotal erythema and edema ensues similar to prolonged testicular torsion (Fig. 2.2), so it is critical to rule out torsion in these patients. Systemic symptoms such as nausea and vomiting are absent with a torsed appendage. The presence of a cremasteric reflex is a helpful physical finding. Generally, the testis is soft in consistency on palpation. A hard palpable, paratesticular nodule representing the torsed appendage in the groove between the superior aspect of the testis and the adjacent epididymis may be present. Additionally, this infarcted appendix testis may demonstrate the "blue dot sign" underneath the scrotal skin.
3. Epididymo-orchitis: Bacterial epididymo-orchitis is rare in children. Bacterial epididymo-orchitis occurs secondary to ascension of a urinary tract infection (UTI) up the vas deferens into the epididymis. Irritative voiding symptoms would be present in setting of a UTI e.g. urinary urgency, frequency and dysuria. The child may have other symptoms such as a fever and the testicle and epididymis are exquisitely tender to touch with variable erythema. When bacterial epididymo-orchitis is present, it may indicate an underlying anatomical abnormality of the genitourinary system. The anatomical abnormality generally results in urinary stasis leading to the UTI. Lower urinary tract dysfunction is another cause for developing a UTI that could lead to epididymo-orchitis. More frequently, there is no evidence of a UTI upon lab testing and this term is often

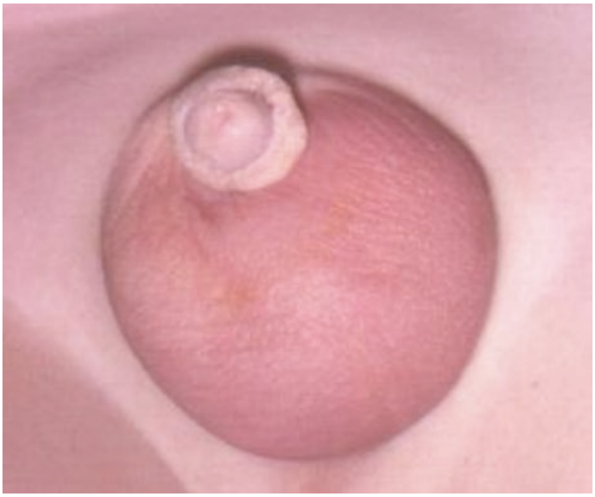

Fig. 2.2 Diffuse left scrotal edema with torsed appendix testis.

applied as a "wastebasket" diagnoses when there is uncertainty of the etiology of the acute scrotum, but is often due to a viral or inflammatory epididymo-orchitis. In a sexually active adolescent, the diagnosis is made after the history, physical exam and laboratory findings are supportive of a sexually transmitted disease.

4. Hernia/Hydrocele: Hernias and hydroceles in children are usually congenital rather than acquired as seen in adults. Pediatric hernias and hydroceles represent a patent processus vaginalis that allows either peritoneal fluid, omentum or bowel to descend into the inguinal canal or scrotum. The history of a fluctuating mass that is reducible on exam is characteristic of the diagnosis. Hydroceles will transilluminate on exam. Hernias and hydroceles are generally not painful; however an incarcerated segment of bowel that becomes trapped or strangulated will be painful and represents a surgical emergency. On rare occasions there have also been reports of diminished blood flow to the testicle identified secondary to an incarcerated hernia, which can also be evaluated at the time of emergent surgery.
5. Spermatic varicocele: Spermatic varicoceles are common in the adolescent population and are frequently discovered during annual physical exams for school or camp. Spermatic varicoceles invariably occur on the left side because of the insertion of the gonadal vein into the left renal vein rather than the vena cava. The majority are asymptomatic however spermatic varicoceles may elicit pain. The pain is usually dull and achy in nature rather than sharp or knife-like. The adolescent will typically complain of a heaviness feeling of the scrotum. Testicular atrophy may be associated with a spermatic varicocele.
6. Spermatocele: Spermatoceles are well circumscribed, round cysts located in the epididymis. Spermatoceles are usually benign and asymptomatic. They are generally located superiorly at the head of the epididymis or inferiorly at the tail of the epididymis. Spermatoceles are frequently discovered during annual physicals for school or camp.
7. Trauma: Most injuries to the testicle(s) are secondary to impact to the scrotum from rough play or sports. The testes are sore and tender to touch. Trauma to the testes may result in a contusion to the testes or rarely, there can be a disruption of the covering of the testes or tunica albuginea.
8. Insect Bite: Insect bites to the scrotum may cause generalized erythema, edema and tenderness to the scrotal skin. There may be a localized, raised papule where the insect bite occurred on the skin.
9. Scrotal Cellulitis: Similar to presentation elsewhere on the body, scrotal cellulitis presents with erythema and edema but is localized to the scrotal wall with no testicular or epididymal involvement.
10. Henoch-Schonlein Purpura (HSP): On rare occasions, this immune-mediated systemic vasculitis has been reported to present in children with acute scrotum, epididymitis, orchitis, and spermatic cord complications, specifically pain, redness and swelling. These symptoms are difficult to link to HSP due to lack of concomitant systemic symptoms, and therefore HSP with scrotal involvement often remains a diagnosis of exclusion.

11. Testicular tumors: A painless, hard testicular mass is presumed cancerous until proven otherwise. Testicular tumors are generally benign in the pre-pubescent child.

Treatment of Abnormalities of the Inguinal Canal and Scrotum

1. Testicular torsion: Any acute and disabling testicular pain should be triaged quickly in the emergency room or office. A quick and expeditious diagnosis is paramount because of the ensuing ischemia. There is a 6 to 8-h window before complete infarction and necrosis of the testes occurs (Fig. 2.3), but salvageability can occur even after this time frame. The diagnosis is typically made from the clinical history and the physical exam, with special scoring systems, such as the TWIST score to aid in diagnosis. The classic history is a post-pubertal child with sudden onset of pain accompanied by nausea and vomiting. The testicle is high-riding and diffusely tender and the child exhibits colicky pain. There is diffuse and increased firmness of the testicle. The cremasteric reflex is absent. A delayed presentation is frequent because the adolescent may not initially disclose their complaints. With a late presentation, the testicle becomes painless and hard. Scrotal sonography is useful in equivocal cases. An initial intervention by manual detorsion under sedation can be attempted depending on the comfort level of the physician. While manual detorsion may decrease ischemia time, it does not obviate the need for surgical treatment. Manual detorsion is typically performed in a medial to lateral rotation however testicular torsion does not always occur in a uniform direction. Referral to a pediatric urologist should be promptly made when the diagnosis of torsion is suspected to plan for emergent exploration and treatment. Treatment is scrotal exploration with

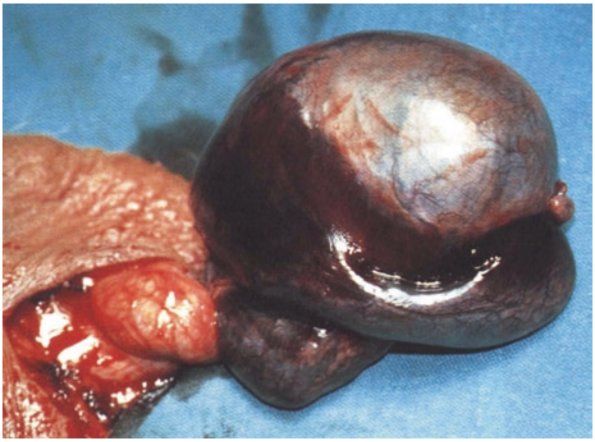

Fig. 2.3 Necrotic testicle secondary to testicular torsion

detorsion of the testes and either orchiopexy vs. orchiectomy with contralateral orchiopexy. Patients with intermittent torsion may have sporadic episodes of severe pain followed by complete relief. Urologic referral is important in suspected cases of intermittent torsion for elective orchiopexy

2. Torsion of the appendix testis or epididymis: Treatment is directed toward symptomatic relief and the resultant inflammation. Patients are advised to take non-steroidal inflammatory drugs (NSAIDs), use scrotal supportive undergarments and to limit physical activity or sports for one week. If symptoms do not improve after 1–2 weeks, or significantly worsen, patients should be instructed to return for further evaluation
3. Epididymo-orchitis: Treatment is directed toward the etiologic cause. If the epididymo-orchitis is idiopathic, then treatment is similar to treatment for torsion of the appendix testis e.g. NSAIDs, scrotal support and limited activity for 7 days. If the etiology is infectious then treatment includes empiric antibiotics until definitive cultures with sensitivity results are available that confirm the empiric treatment or direct new and different antibiotics. If there is suspicion that the patient has a sexually transmitted disease, the adolescent is treated with Ceftriaxone 250 mg intramuscular in a single dose plus Doxycycline 100 mg orally twice a day for 10 days. For acute epididymitis most likely caused by enteric organisms or with negative gonococcal culture or nucleic acid amplification test, treatment is Ofloxacin 300 mg orally twice a day for 10 days or Levofloxacin 500 mg orally once daily for 10 days.
4. Hernia/Hydrocele: Treatment for hernias and hydroceles is surgical. If the hernia is reducible, then surgical treatment may be scheduled electively as an outpatient procedure. If the hernia is non-reducible with strangulation of a segment of bowel, then emergent surgical intervention is warranted.
5. Spermatic varicocele: The indications for treatment for varicoceles in an adolescent are different than in an adult. Fertility issues are the primary indication for treatment of varicoceles in adults and are not applicable to adolescents. The indications for treatment for adolescents include (1) scrotal or testicular pain or (2) testicular atrophy of the affected side. Another consideration for treatment would include an extremely large spermatic varicocele such that the child is self-conscious and has self-esteem issues. The treatment for spermatic varicoceles is either through ligation/division of the spermatic veins or embolization of the spermatic veins.
6. Spermatocele: Spermatoceles are typically innocuous and pose no concern. Observation is the usual course. Rarely, spermatoceles may become quite large and elicit pain or become quite unsightly. If so, then surgical excision is reasonable.
7. Trauma: The treatment for scrotal trauma is dependent upon the underlying resultant injury. If there is testicular or scrotal wall contusion, then treatment is supportive care with NSAIDs, scrotal support and limited physical activity. If there is a testicular rupture of the testes, then emergent scrotal exploration with testicular debridement and closure is warranted. Diagnosis of testicular rupture

is often difficult on ultrasound, so clinical suspicion may sometimes guide decision to proceed to the operating room

8. Insect Bite: Treatment is no different than for insect bites at other skin sites. For scrotal edema, treatment is supportive care with NSAIDs, scrotal support and limited physical activity.
9. Scrotal Cellulitis: Treatment is similar to treatment of cellulitis located elsewhere in the body with antibiotics and supportive care. If exam is limited due to pain, ultrasound may be necessary to rule out any depper involvement of the testes, epididymis or spermatic cord.
10. Henoch-Schonlein Purpura (HSP): Unless there is concomitant HSP presentation with purpura or lesions systemically, it is difficult to diagnosis HSP of the scrotum. Treatment is generally supportive with rest, hydration and symptom relief. NSAIDS can be used and a report of low molecular weight heparin for spermatic vein thrombosis
11. Testicular tumors: Treatment is dependent on staging. Staging workup includes serum testing with a complete blood count, complete metabolic panel including a hepatic panel, lactic dehydrogenase, alpha fetoprotein level and beta human chorionic gonadotropin level. Radiologic imaging includes computerized tomography (CT) of the chest, abdomen and pelvis or a chest x-ray with a CT of the abdomen and pelvis. Ultrasonography may be helpful in the surgical planning of pre-pubertal testes tumors which are more likely to be benign and may be amenable to a partial orchiectomy in select cases.

Indications for Referral

1. Testicular torsion: Once the diagnosis is suspected, referral to a pediatric urologist is mandatory.
2. Torsion of the appendix testis or epididymis: Referral is dependent on the comfort level of the primary care provider with the diagnosis.
3. Epididymo-orchitis: If an anatomical congenital anomaly of the urinary tract is found, then a referral to a pediatric urologist is warranted.
4. Hernia/Hydrocele: Referral to a pediatric urologist or surgeon is warranted after the diagnosis is made.
5. Spermatic varicocele: Referral to a pediatric urologist or interventional radiologist is warranted if there is an indication for treatment as listed above.
6. Spermatocele: Referral is not necessary if the patient is asymptomatic.
7. Trauma: Referral is dependent on the degree of injury. Contusions can be managed by the primary care provider, but if there is concern for testicular rupture emergent referral is needed.
8. Insect Bite: Referral is generally not necessary.
9. Scrotal cellulitis: Referral is no necessary unless there is concern for additional urologic organ involvement

10. H Henoch-Schonlein Purpura (HSP): Referral is not necessary unless there is concern for other urologic diagnoses
11. Testicular tumors: Referral to a pediatric urologist and an oncologist is warranted once the diagnosis is suspected.

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Chapter 3
Approach to Scrotal Abnormalities

Daniel T. Keefe, Fardod O'Kelly, and Martin Koyle

Key Points
- Diagnosing scrotal pathology is often accomplished with history and physical exam
- Ultrasound can be used in cases of diagnostic uncertainty
- Testicular torsion is a urologic emergency that requires urgent specialist referral and surgical exploration
- A painless testicular mass is a testicular tumour until proven otherwise

Introduction

The acute scrotum is the development of pain and/or swelling of the intrascrotal contents. It is a common condition in the pediatric population and can represent a wide variety of clinical pathology. There is a broad differential diagnosis with varying levels of acuity (see Table 3.1). The diagnosis can often be made with history and physical examination alone; however, laboratory tests and diagnostic imaging can be used as adjunctive diagnostic tools. This chapter will review common causes of an acute scrotum in children and adolescents and assist the primary care provider in making the diagnosis and initiating appropriate management.

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Table 3.1 Differential diagnosis of scrotal pathology with typical clinical and radiographic findings and appropriate initial management

Differential diagnosis	Typical presentation	Examination findings	Ultrasound findings	Management
Testicular Torsion	• Acute onset severe pain • Nausea/vomiting	• Diffusely tender, hard testicle • Horizontal lie of testicle • Absent cremasteric reflex	• Absent blood flow • Twisted spermatic cord	• Emergent Urology referral for surgical exploration orchiopexy vs orchiectomy AND contralateral orchiopexy • Testicular salvage rates (time from symptom onset): [3] <6 h: 90–100% >12 h: 50% >24 h: <10%
Appendix testis/epididymis torsion	• Gradual to acute onset of testicular pain • Absent systemic symptoms • Localized testicular pain	• Tender localized palpable mass • Otherwise normal soft testicle • Presence of cremasteric reflex • "Blue dot sign"	• Normal blood flow • Hypoechoic torsed appendage	• Conservative/symptomatic management • NSAIDs, scrotal support and activity reduction
Acute epididymitis/orchitis	• Lower urinary tract symptoms • Fever • Acute/subacute testicular pain	• Scrotal edema/erythema • Normal testicular lie	• Increased blood flow to epididymis or testicle	• Treatment based on etiology • *Non-infectious*: conservative management (as above) • *Infectious*: empiric antibiotics and await culture sensitivities • *Sexually active adolescents*: Ceftriaxone 250 mg IM × 1 dose + Doxycycline 100 mg PO BID × 7 days *OR* Azithromycin 1 g PO × 1 dose *Children/Negative STI cultures*: Ofloxacin 300 mg PO BID × 10 days *OR* Levofloxacin 500 mg PO BID × 10 days

Table 3.1 (continued)

Differential diagnosis	Typical presentation	Examination findings	Ultrasound findings	Management
Hernia/ Hydrocele	• Painless fluctuating scrotal mass	• Reducible hemiscrotal swelling • Transillumination identifying cystic fluid collection	• Normal blood flow • Hemiscrotal fluid collection	• Initial observation • *Communicating hydrocele* persisting >1 year of age: Refer for elective surgical intervention • *Non-Communicating hydrocele*: Refer for elective surgical intervention if symptomatic or for cosmesis • *Incarcerated hernia*: urgent surgical referral
Trauma	• Impact to the scrotum from rough play or sports • Traddle injury	• Diffusely painful testicle • Visible hematoma	• Parenchymal fracture or tunica albuginea rupture • Hematocele	• *Testicle contusion*: supportive management • *Testicular fracture/ rupture*: urgent scrotal exploration with debridement and repair
Varicocele	• Asymptomatic to dull ache • Prominent scrotal veins	• 90% left side • Venous distention while standing or on Valsalva • "Bag of worms" sign	• Dilated, tortuous pampiniform plexus	• Observation • Consider referral for surgical treatment if: symptomatic, ipsilateral testicular hypotrophy (>20% or >2mL on US), fertility concerns
Spermatocele	• Painless scrotal swelling	• Well-circumscribed round cysts located at the epididymis	• Fluid filled cyst attached to the epididymis	• Observation • Consider referral if symptomatic or patient desire to surgical excision
Testicular Tumour	• Painless testicular mass	• Palpable testicular mass	• Solid testicular mass	• Urgent referral to Urologist for *radical inguinal orchiectomy*

History

The patient's history should be fully elucidated. When the child presents with scrotal pain it is important to identify the *pain onset*, *severity*, *duration*, *prior episodes* and any other associated features including *lower urinary tract symptoms* (e.g., dysuria, frequency, hematuria, urethral discharge), *nausea/vomiting*, *fever*, *sick contacts*, and *prior illnesses*. A history of testicular *trauma* can point toward a testicular contusion, or, uncommonly, a testicular fracture caused by disruption of the tunica albuginea.

Acute onset of severe testicular pain, often associated with nausea and vomiting can point more toward *testicular torsion* as the source of pain. Often patients will report prior episodes of acute pain which resolve spontaneously indicating torsion-detorsion phenomenon. *Appendix testis/epididymis torsion* may mimic similar symptoms, but pain is often in a specific location (e.g., superior pole of the testicle).

Pain associated with infectious signs or symptoms (e.g., fever, lower urinary tract symptoms) most likely represents *acute epididymitis/orchitis*. A thorough *sexual history* should be completed when appropriate to identify risk factors for sexually transmitted infections. Prior urologic surgeries or genitourinary infections can identify patient's that may have an abnormal genitourinary tract predisposing to recurrent infections.

Painless swelling of the scrotum that fluctuates in size generally represents a communicating *hydrocele* due to peritoneal fluid traversing through a patent processus vaginalis. This is a form of *inguinal hernia* and therefore loops of bowel can become incarcerated and lead to abdominal pain and gastrointestinal symptoms. A *non-communicating hydrocele* is usually found in older children and adolescents and can be idiopathic or secondary to an inflammatory/reactive process whereby fluid gets secreted from the mesothelial lining of the tunica vaginalis. It is contained within the tunica vaginalis and therefore does not fluctuate in size.

A *painless testicular mass* identified on self or clinical exam is a *testicular tumour* until proven otherwise. It is important to identify symptoms that may represent metastatic disease including neurologic symptoms, abdominal masses or discomfort, or respiratory symptoms.

Physical Examination

A generalized physical examination should be completed making note of the patient's *generalized appearance*. Assess *vital signs* particularly for evidence of infection (e.g., fever, tachycardia).

Abdominal exam should be completed to identify any overt *abdominal masses* or pain. Pain from *testicular torsion* can radiate into the inguinal canal. Abdominal pathology can also manifest as *referred pain* into the scrotum, for example, in the case of a *distal ureteric calculus*.

Genitourinary exam should assess overall appearance. Take note of the general appearance of the scrotum including any *erythema*, *edema*, and *swelling*. Assess for a *high riding testicle* or *horizontal lie of the testicle* (Fig. 3.1a). Attempt to elicit a *cremasteric reflex*. On palpation assess for diffuse vs localized tenderness of the testicle. Assess for any *palpable masses*. Comment on the testicular *consistency* (soft vs hard). If a hydrocele is present is it reducible or non-reducible? If there is a varicocele, is it bilateral vs unilateral? Is the varicocele present when standing and supine? Left are more common (Fig. 3.1b) (~90%). A varicocele on the right warrants further investigation to assess for inferior vena caval compression [1].

Adjunct physical exam findings:

- *Cremasteric reflex*: This is elicited by lightly stroking the inner portion of thigh and assessing for testicular ascent. Absent cremasteric reflex can support the diagnosis of testicular torsion
- *Blue dot sign*: This is a pathognomonic finding of appendix testis torsion if identified. It is a blue discolouration noted beneath the scrotal skin and is found in ~20% of patients with appendix testis torsion (Fig. 3.1c) [2].
- *Transillumination*: illuminate hemiscrotal swelling to assess for a hydrocele (Fig. 3.1d).

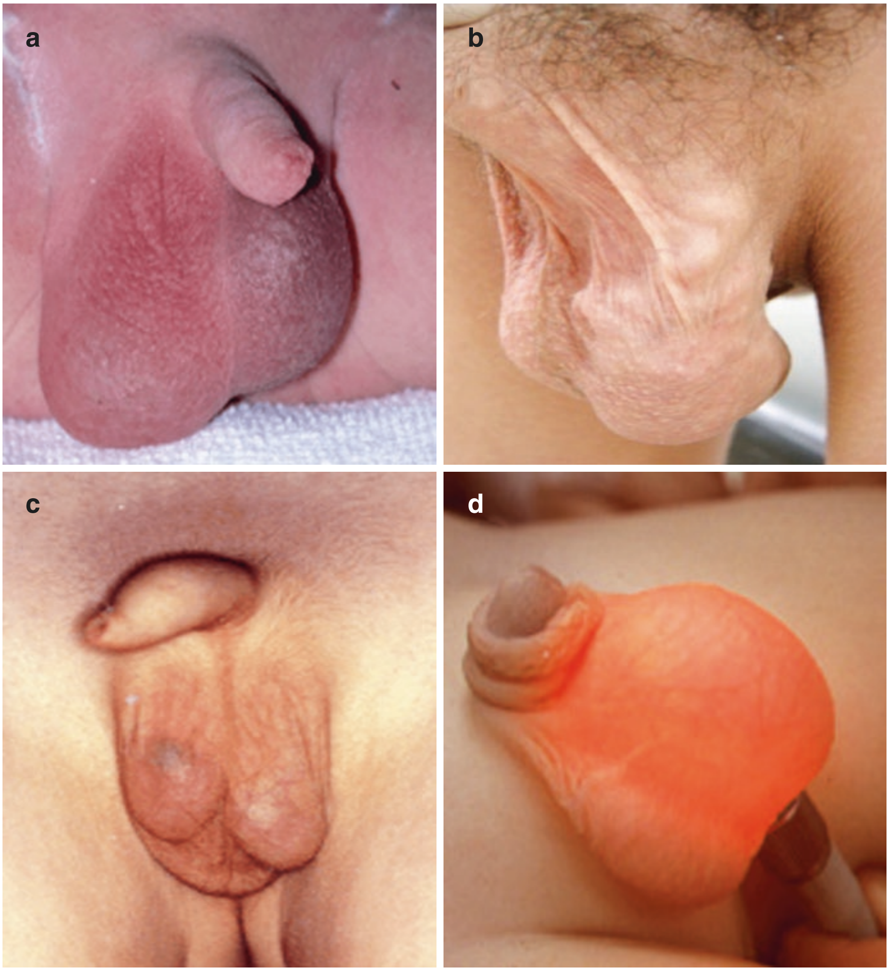

Fig. 3.1 (**a**) High riding left testicle in a patient with testicular torsion. (**b**) 'Bag of worms' sign in a patient with a left varicocele. (**c**) Right sided appendix testis torsion with associated 'blue dot sign'. (**d**) Transillumination of a left sided hydrocele

Laboratory Tests

Laboratory investigations are not mandatory for most conditions however can be beneficial in certain situations.

Suspicion of testicular malignancy:

- Serum tumour markers: beta subunit of human chorionic gonadotropin (beta-HCG), alpha-fetoprotein (AFP) and lactate dehydrogenase (LDH)

Suspicion of epididymitis or orchitis:

- Urine microscopy and culture
- Culture for sexually transmitted infections (*N gonorrhea* and *C trachomatis*)

Diagnostic Imaging

Diagnostic imaging tests can be used in situations of diagnostic uncertainty however, in most cases, history and physical exam is adequate in making the diagnosis for scrotal pathology.

Colour Doppler Scrotal Ultrasonography can distinguish between *torsion* and *epididymitis/orchitis*. In the setting of scrotal trauma, ultrasound can help identify *testicular fracture* or rupture of the tunica albuginea. It is also important for radiographic identification of suspected *testicular tumours* or if there is concern for a pathologic *varicocele*.

Testicular tumour requires the following further imaging to rule out metastases:

- Chest X-ray or CT chest
- CT abdomen and pelvis
- CT head if neurologic symptoms

Management

Testicular Torsion

- Suspected testicular torsion should be triaged urgently. When there is adequate clinical suspicion of testicular torsion immediate Urology referral should be obtained.
- Clinical diagnosis can be aided by the use of decision tools. Barbosa et al. [5] developed the *TWIST score* (Testicular Workup for Ischemia and Suspected Torsion) which was validated for use by non-urology medical personnel in 2016. This can also facilitate clinical discussion with surgical specialists to facilitate expedited care to those at highest risk of testicular torsion [4, 5].

TWIST Score

Clinical finding	Point(s)
Testis swelling	2
Hard testis	2
Absent cremasteric reflex	1
Nausea/vomiting	1
High riding testis	1
Total	*7 points*

Risk category	TWIST score	Recommendation
Low risk	0	Observation (no testicular torsion)
Intermediate risk	1–5	Scrotal ultrasound
High risk	≥6	Tanner stage 1–2: Scrotal ultrasound Tanner stage 3–5: Surgical exploration

- Classically, there is an approximate 6-h window from symptom onset in order to salvage the testicle before complete infarction and necrosis set in. Intraoperatively, the surgeon will assess testicular viability. If the testicle appears viable a *bilateral orchiopexy* will be completed as there is a high risk of contralateral testicular torsion. This is due to the main pre-disposing anatomic variant known as the *bell-clapper deformity* which is commonly present bilaterally. If the testicle appears to be non-viable an orchiectomy should be performed in addition to the contralateral orchiopexy (Fig. 3.2) [6].

Appendix Testis/Epididymis Torsion

- Treatment is primarily to manage symptoms and relieve swelling. General recommendations include NSAIDs, scrotal supportive undergarments and reduced activity for the following 1–2 weeks. [7]

Acute Epididymitis/Orchitis

- Treatment is dependent on the suspected underlying etiology. Empiric antibiotics are recommended until final culture results are available. Generally, patients are safely managed in an outpatient setting however if there is any sign of associated

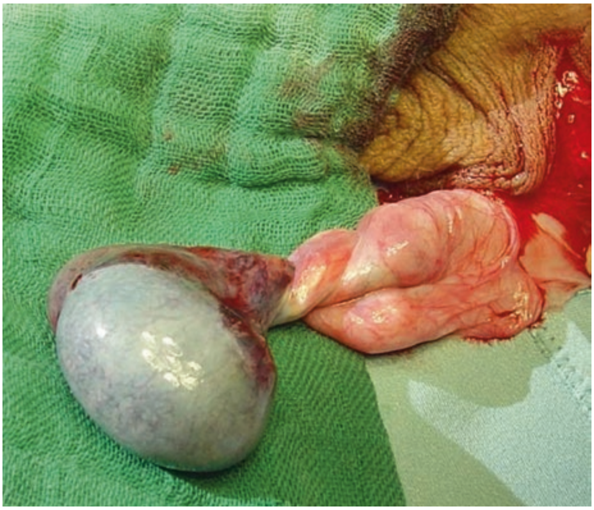

Fig. 3.2 Intraoperative findings of an acute testicular torsion

sepsis the patient should be admitted for monitoring and parenteral antibiotics. Epididymitis/orchitis are inflammatory conditions of scrotum which can be infectious or non-infectious. In younger children, the most common cause relates to enteric organisms. In sexually active adolescents the most common organisms are *Chlamydia trachomatis* and *Neisseria gonorrhea*.

- Non-infectious etiology suspected:
  - Symptomatic management: NSAIDs, scrotal supportive undergarments and reduced activity
- Infectious etiology suspected:
  - *Sexually active adolescents*: Ceftriaxone 250 mg IM × 1 dose + Doxycycline 100 mg PO BID x 7 days *OR* Azithromycin 1 g PO x 1 dose
  - *Children/Negative STI cultures*: Ofloxacin 300 mg PO BID × 10 days *OR* Levofloxacin 500 mg PO BID × 10 days [8]

Hernia/Hydrocele

- Communicating hydroceles are more common in infants in the first year of life and often resolve spontaneously. Therefore, an initial period of observation is recommended. If the hydrocele persists beyond 1 year of age surgical management can be considered and referral to a urologist should be obtained [9].
- Non-communicating hydroceles often arise secondary to underlying inflammatory pathology and thus if you are unable to reliably examine the testicle a scrotal ultrasound should be obtained. If underlying acute pathology is ruled out, consideration for surgical hydrocelectomy can be considered if the hydrocele is symptomatic or the patient desires surgery for cosmetic purposes
- Hernias that are reducible can be referred for elective surgery however if there is an incarcerated hernia the patient should be assessed urgently by a surgeon.

Trauma

- Scrotal trauma often arises from blunt trauma (e.g., sports, falls, straddle injuries). Penetrating trauma is exceedingly rare but should prompt urgent surgical assessment and exploration. Clinical exam may identify signs of a hematocele/hematoma making examination of the testicle challenging. Doppler ultrasound can be used to identify testicular fracture or rupture of the tunica albuginea
- Testicular contusion without fracture/rupture can be treated conservatively with symptomatic management
- Testicular fracture/tunica albuginea rupture warrants urgent urology referral and scrotal exploration with debridement of necrotic tissue and repair (Fig. 3.3) [10]

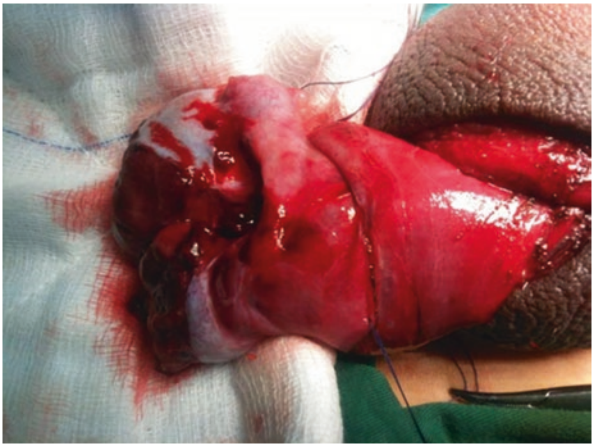

Fig. 3.3 Intraoperative findings of a testicular trauma with disruption of tunica albuginea

Varicocele

- Varicoceles are common and concerns for fertility potential when they are identified is often a major concern for patients and their families. The main indication for surgery in adults is infertility. This is less applicable in adolescents. The primary indications for surgery in adolescents include: (1) persistent symptoms and (2) ipsilateral testicular hypotrophy (usually >20% size discrepancy on exam or >2 mL volume difference on ultrasound) [11]. Treatment would include ligation/division of spermatic veins or embolization [12].

Spermatocele

- This is a benign fluid filled cyst and therefore in most cases observation is recommended
- If they become large, symptomatic or there is concern for cosmetic appearance then referral to urology for consideration of elective spermatocelectomy can be considered

Testicular Tumour

- If a testicular tumour is identified urgent urology referral should be completed. This will permit appropriate staging via laboratory investigations (serum tumour markers with AFP, bHCG and LDH), chest imaging, and CT abdomen/pelvis. Staging and underlying pathology identified on radical inguinal orchiectomy will guide further treatment.

Conclusion

Scrotal abnormalities represent a host of underlying pathology with varying levels of acuity. Most pathology can be discerned primarily on history and physical examination however, if there is any diagnostic uncertainty a scrotal Doppler ultrasound can be obtained. Management will depend on the underlying pathology. Testicular torsion and testicular tumours are urgent conditions that need urgent urology referral. The remaining differential diagnoses can often be managed empirically with elective referral to urology on a case by case basis depending on complexity or health care provider's level of comfort [13].

Acknowledgments All photos were retrieved from Google.

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Chapter 4
Disorders of Male External Genitalia: Problems of the Penis and Foreskin

Prasad P. Godbole

Key Points

- A non retractile foreskin at birth is normal. Spontaneous retractility begins anytime between 2 and 3 years of age. Ballooning of the foreskin is normal as are occasional episodes of posthitis
- Majority of boys will have a retractile foreskin by 10 years of age and 95% by 16–17 years of age.
- The only absolute medical indication for circumcision is penile malignancy and balanitis xerotica obliterans.
- Most inflammatory conditions of the foreskin can be managed conservatively in primary care and do not need referral.

Introduction

The management of foreskin conditions varies amongst medical practitioners from observation to circumcision. In the authors experience, poor understanding of the natural history of the foreskin has led to an increasing number of referrals into specialist centres for evaluation and/or circumcision. This can lead to a huge strain on resources when many of these conditions can be successfully be managed in primary care. This chapter deals with common foreskin problems, their etiology and

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management in primary care. Indications for referral will be highlighted. Circumcision will be dealt with in another chapter.

Common Foreskin Conditions

1. Non retractile foreskin: Almost all boys have a non retractile foreskin at birth. The inner foreskin is attached to the glans. Foreskin adhesions break down and form smegma pearls presenting as yellowish white cysts under the fore-skin which are then extruded. Foreskin retractility begins after 2 years of age. This process is spontaneous and does not require manipulation. A non retractile foreskin on gentle attempted retraction pouts like a flower-physiological phimosis (Fig. 4.1). The majority of boys will have a retractile foreskin by 10 years of age and 95% by 16–17 years of age.
2. Balanoposthitis: inflammation of the glans and the foreskin.
3. Balanitis: inflammation of the glans that often spreads along the shaft and may occur in the circumcised population.
4. Posthitis: inflammation restricted to the foreskin itself.
5. Balanitis Xerotica Obliterans(BXO): a lesion akin to lichen sclerosus et atrophicus. This causes true phimosis—pathological phimosis. This causes a shutter type foreskin with no pouting of the inner foreskin on gentle retraction (Fig. 4.2). It is rare before 5 years of age.
6. Paraphimosis: results when the narrow tip of the foreskin is retracted behind the glans at the coronal sulcus causing edema of the glans and foreskin and inability to manipulate the foreskin back over the glans (Fig. 4.3).

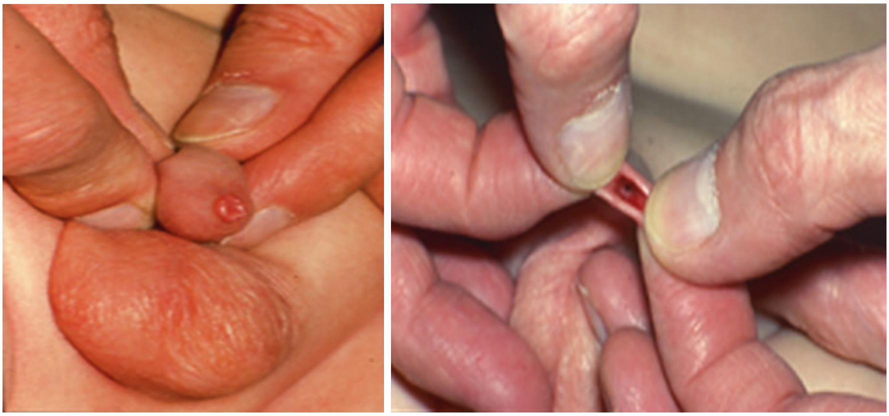
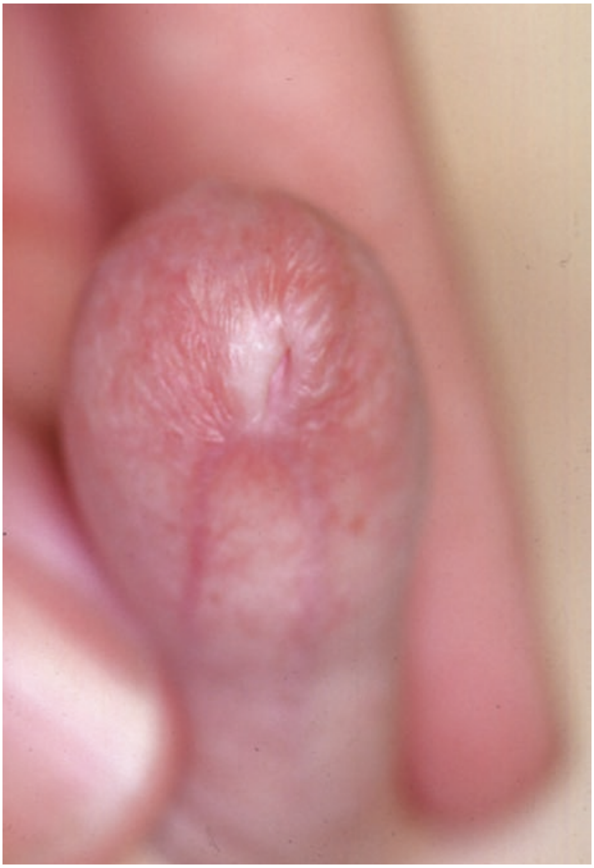


Fig. 4.1 Healthy non retractile foreskin (physiological phimosis). Inner layer pouts out like a flower

Fig. 4.2 Balanitis Xerotica Obliterans. Note the rigid prepuce that cannot be retracted and does not pout like a healthy non retractile foreskin as in Fig. 4.1



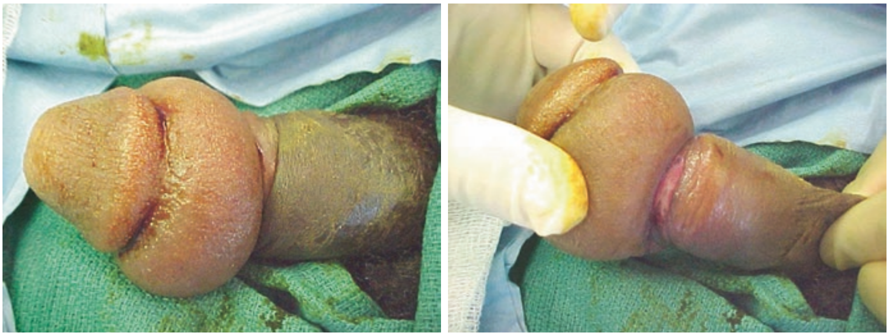


Fig. 4.3 Paraphimosis: note the constricting band at the coronal sulcus

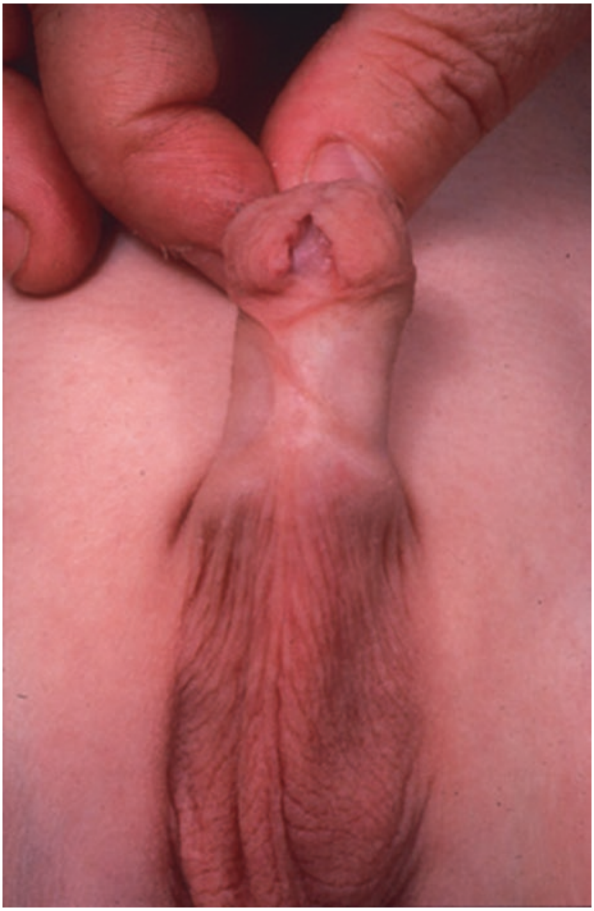


Fig. 4.4 Hooded foreskin

7. Hooded foreskin: is an abnormal dorsal hemiforeskin which is deficient ventrally and is usually associated with hypospadias (Fig. 4.4). Hypospadias will usually be identified on the newborn check but minor variants may be missed and present later in infants.
8. Localised collection of smegma: Due to the non retractile foreskin, desquamated epithelial cells from the inner layer of the foreskin collect around the coronal sulcus and can appear as an oval yellowish lump (Fig. 4.5). This often causes anxiety amongst parents and primary care practitioners alike. This lump is known as smegma pearls and is entirely normal and requires no intervention.
9. Buried penis : In many cases, boys are referred to specialists with concerns about the size of the penis. In the majority the boys, the penis retracts in a large suprapubic fat pad thereby giving it a shorter appearance. On displacing this fat pad, the true length of the penile shaft can be demonstrated. This does not require any intervention. Megaprepuce or a volcano penis (Fig. 4.6) is the term given when there is redundant inner prepuce and abnormal dartos attachments

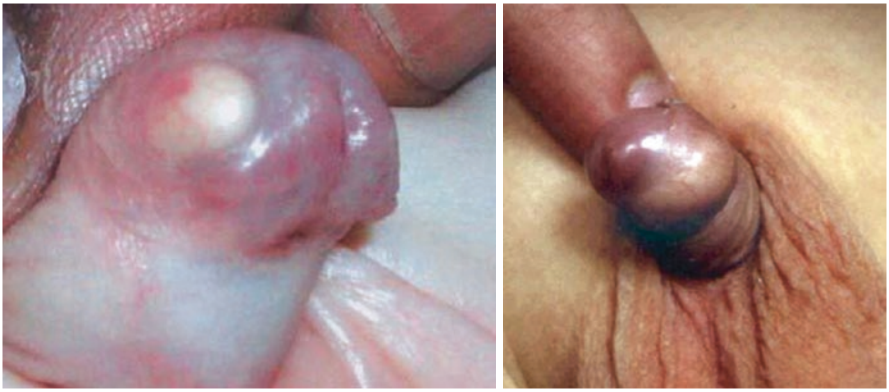
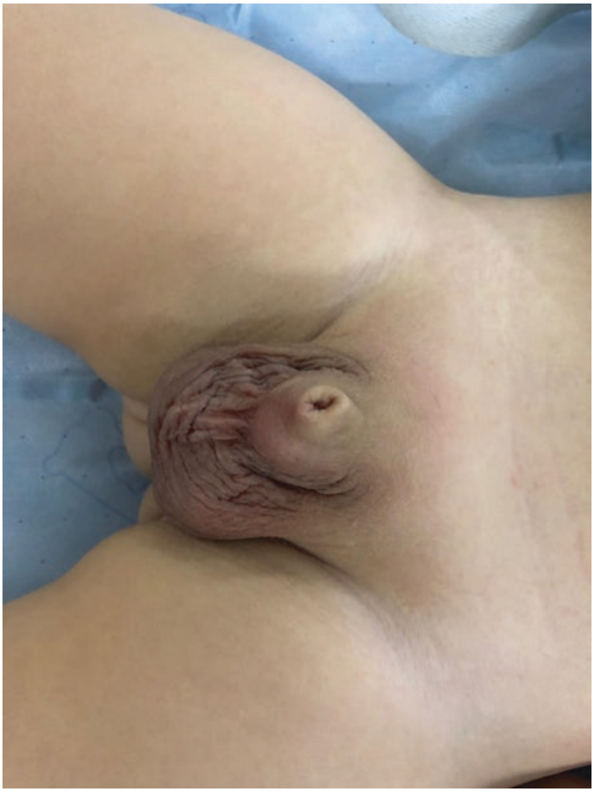


Fig. 4.5 Localised collection of smegma: note the non retractile/partially retractile foreskin within which the smegma collects

Fig. 4.6 Buried (volcano penis)



which tethers the penis down. When boys void, this leads to a golf ball size ballooning of this redundant prepuce from which parents have to milk the urine out. This requires surgical intervention for the more severe forms of the condition.

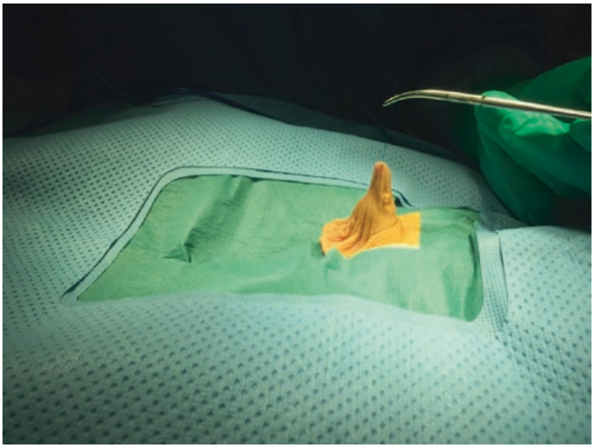


Fig. 4.7 Penoscrotal web: note the scrotal encroachment on the penile shaft

10. Penoscrotal web : In this condition there is encroachment of the scrotal skin onto the shaft of the penis causing ventral skin tethering and curvature of the penis (Fig. 4.7). In milder cases, the scrotal skin drops back into a normal position and does not requires treatment. In some cases surgical intervention is required.

Treatment of Conditions of the Foreskin

1. Non retractile foreskin: This does not require any treatment if the foreskin is healthy. Topical steroids are known to hasten retractility of the foreskin and may be considered. On no account should the parents be asked to forcibly retract the foreskin. In older pubertal children with a healthy non retractile foreskin, a preputioplasty may be considered.
2. Localised collection of smegma pearls: No intervention is necessary.
3. Inflammatory conditions: balanoposthitis, balanitis, posthitis: Simple bathing, topical steroids, and antibiotics. If recurrent disabling attacks of balanoposthitis occur despite conservative management, a circumcision may be considered.
4. Balanitis Xerotica Obliterans: Circumcision.
5. Paraphimosis: Reduction with or without an anesthetic.
6. Hooded foreskin: without hypospadias: no treatment, modified circumcision or foreskin reconstruction. If with hypospadias: no treatment, modified circumcision or fore-skin reconstruction with hypospadias repair.
7. Buried penis: requires surgical intervention to release the abnormal attachments and excise the redundant inner prepuce with a modified circumcision. No intervention where the buried penis is purely due to a large suprapubic fat pad.
8. Penoscrotal web: minor webbing does not require surgery. Significant tethering will require surgical intervention.

Indications for Referral

1. Recurrent severe balanoposthitis where conservative management has not been successful.
2. Balanitis xerotica obliterans.
3. Paraphimosis where reduction is not possible with simple manual reduction.
4. Hooded foreskin with or without hypospadias: If parents wish surgery for cosmetic/functional reasons.
5. Significant buried penis with symptoms
6. Penoscrotal web with functional problems

Suggested Reading

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Chapter 5
Disorders of Male External Genitalia: Hypospadias, Epispadias, Concealed Penis, Urethral Disorders

Abraham Cherian

Hypospadias

Key Points

- It is a developmental anomaly of the penis usually diagnosed at birth and is comprised of three key components of varying degrees namely a hooded foreskin, curvature of the shaft (chordee) and a urethral opening falling short of the tip of the penis. Severity depends on the location of the opening, quality of the ventral spongiosum proximal to the meatus and the curvature.
- Circumcision is not recommended until a specialist has been consulted.
- Most commonly occur in isolation but can be associated with undescended testes in a minority. When associated with undescended testes a referral to a multidisciplinary team that deals with disorders of sex development should be made.
- Uncommonly it is diagnosed when a child is scheduled for circumcision and when the foreskin is retracted a sub-terminal urethral opening is noted. The procedure should be abandoned and referred to a specialist.

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- Not all need surgery and increasingly the milder forms when the opening is well within the glans and without a curvature are managed conservatively.
- Penile curvature and misdirected urinary stream usually need surgical correction
- Although the minority is severe and requires a two-stage operation, most are less severe and require a single stage operation around a year of age.
- The most commonly encountered complications of surgery include urethro-cutaneous fistula and meatal stenosis. Less commonly urethral stricture, diverticulum and recurrent penile curvature in the long term are reported.

Introduction

Hypospadias is one of the commonest conditions seen in pediatric urology practice. Genetic and environmental factors play a significant role in its etiology. There is a wide variation in the prevalence reported from a variety of studies however most report between 5 and 50 per 10,000 births [1]. Prematurity does not increase the risk, as penile development is complete by 16 weeks of gestation. Most are diagnosed at birth and some when circumcision is attempted. In the majority the hooded foreskin reveals the abnormality however in some the prepuce may be intact and can hide the abnormal meatus, which is usually quite large. This variant is referred to as a mega-meatus intact prepuce (MIP; Fig. 5.1a, b). In both scenarios, circumcision should not be performed and appropriate referral to a specialist is recommended. Penile curvature is variable and although quite obvious in the severe or ones with a more proximal meatus, it is less easily appreciated in the more distal ones especially in the non-erect penis (Fig. 5.2). At times having a distal meatus may be interpreted as a less severe anomaly however the quality of the ventral spongiosum may be poor

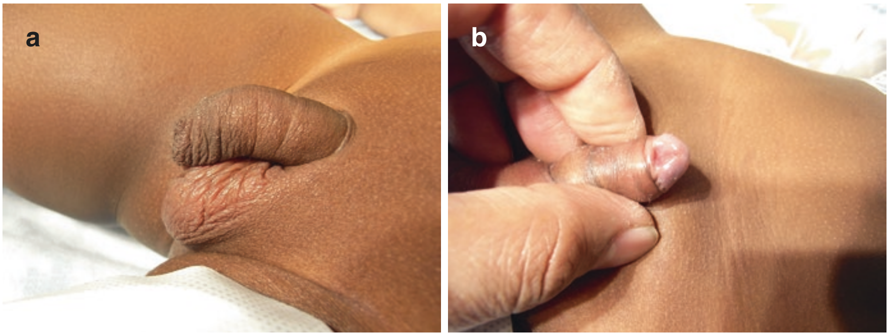


Fig. 5.1 (**a**) Foreskin covering the entire glans in MIP. (**b**) Foreskin retracted revealing megameatus in MIP

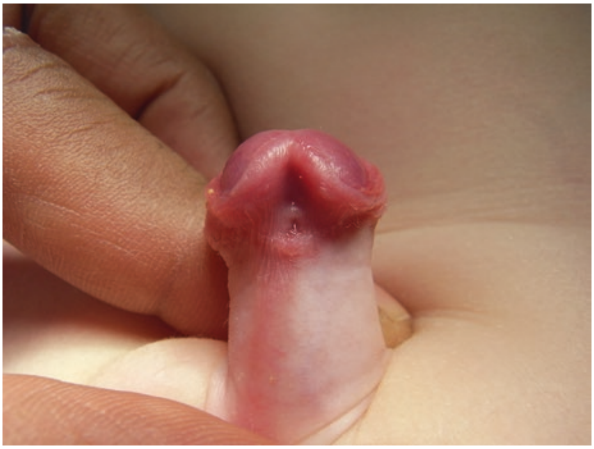


Fig. 5.2 Distal hypospadias with no obvious curvature

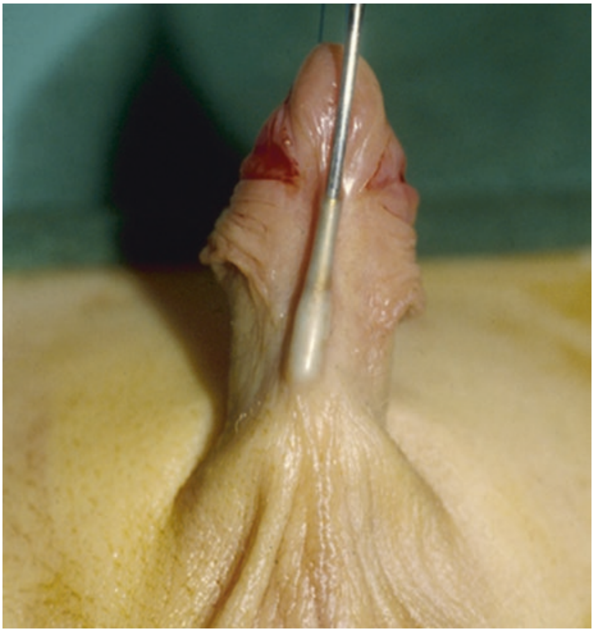


Fig. 5.3 Distal meatus but deficient distal spongiosum up to mid-shaft

and this is evidenced by a very thin urethral segment until the proximal end of the spongiosal divarication (Fig. 5.3). Often this will need a two-stage operation and should not be attempted by the less experienced.

Associated anomalies are seen in fewer than 15% of hypospadias and include undescended testis, hernia, and hydrocele. It is rarely associated with upper urinary tract anomalies and hence ultrasound examination is not recommended routinely. The more severe forms may be associated on the rare occasion with a prostatic utricle.

This may make catheterization challenging at the time of repair. When associated with undescended testis referral to a multidisciplinary team dealing with disorders of sex development is imperative.

Management

All boys with hypospadias or suspected hypospadias, hooded foreskin, penile chordee, and megameatus should be referred to a pediatric urologist when the diagnosis is made. In most, the indication to intervene is the potential to cause a misdirected stream and penile curvature. In the absence of both and especially with the meatus on the glans, we are increasingly seeing a trend to manage them conservatively. The timing of intervention is variable but most tend to operate between 1 and 2 years of age and some prefer in infancy [2]. Children seem to fare better both emotionally and psychologically within this time frame although surgical outcomes are no different when done at a later date. The goal of surgery is to achieve a terminal and adequate meatus on the glans, correct the penile curvature along with a good cosmetic outcome. In the majority a single stage operation is adequate to achieve the above, however, in the minority, which is severe, this can only be achieved in 2 stages (Fig. 5.4). This is usually done if the urethral plate needs transection to correct severe chordee. Most centers would perform this as a day case surgery but some of the more severe ones are kept overnight in hospital. In the two-stage operation, the first instance involves the correction of curvature with grafting of the defect using the foreskin and the second stage involves tabularization of this grafted area (Fig. 5.5a, b). Catheters or stents are usually employed during these procedures between 5 and 10 days. Perioperative testosterone is used by some especially if the glans or shaft is small, to facilitate growth and repair. This is given as two doses of

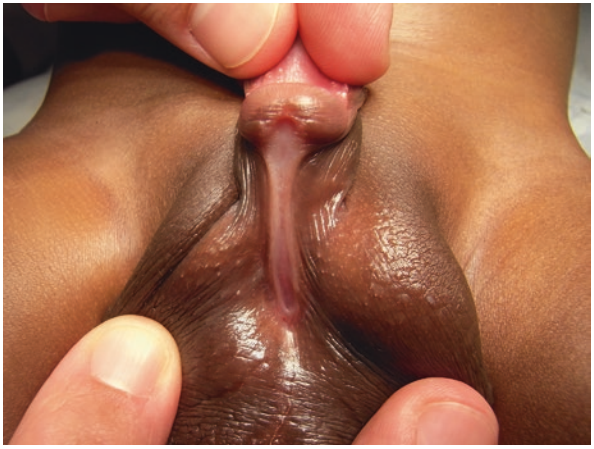


Fig. 5.4 Proximal hypospadias. Note meatus at scrotal or perineal level with severe chordee

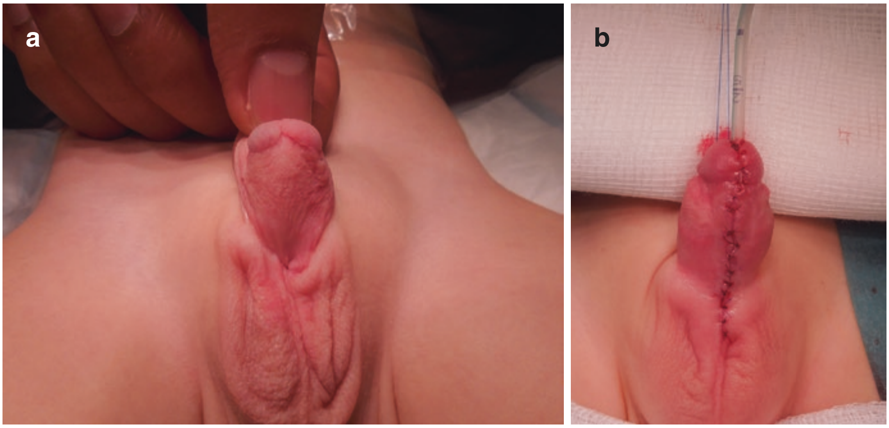


Fig. 5.5 (**a**) Two-stage repair—6 months following First stage repair with preputial graft. Note meatus is at the peno-scrotal junction. (**b**) Two stage repair—second stage completed wherein the grafted plate is tubularized over a stent

intramuscular testosterone a month apart and the repair scheduled a month after the last dose. Some controversy exists regarding testosterone with reports suggesting a complication rate with its use.

Complications

The most common complication in hypospadias surgery is a urethro-cutaneous fistula, which may not become apparent until they are out of nappies evidenced by more than one urinary stream. Typically it is a small drip or fine stream at an unexpected location other than the terminal meatus. Some fistulas close spontaneously however they will persist if there is a distal narrowing. Fistulas that do not resolve by 6 months will usually need formal closure and exclusion of distal narrowing.

Meatal and or urethral narrowing presents with a thin or fine stream, new onset wetting, dribbling, urinary tract infection, straining and rarely epididymo-orchitis. Prompt referral is important, as this will need treatment either in the form of a meatotomy or meatoplasty. Meatal dilatation alone may not yield a good outcome in the long term and may entail several interventions. The more proximal strictures and resistant stenosis may require a laying open of the previous repair followed by excision of scar tissue, grafting and then a redo repair in a staged manner.

Recurrent or persistent penile curvature may be noticed by parents or presents at puberty. Generally, curvatures beyond 30 degrees will need re-intervention, as these may impair sexual function.

Urethral diverticulum although rare, is more commonly encountered with proximal types of hypospadias repair. A ventral bulge during the act of voiding and manual compression of the bulge yielding urine following the act is characteristic.

These can be managed conservatively but larger ones and those with abnormal voiding pattern may need excision and repair.

Distal glans breakdown or dehiscence leading to meatal regression is probably under-reported. More severe glans or urethral breakdown is fortunately rare and redo surgery will need careful discussion.

Epispadias

Key Points

- Epispadias is a rare penile abnormality wherein there is a dorsal midline defect of varying severity affecting glans only (glanular) or glans and shaft (penile) or more proximally (pubic/penopubic). This results in a wide dorsal opening of the urethral meatus sometimes concealed by an intact foreskin in the distal glans only types but is more obvious in the penile and more proximal defects where the foreskin is deficient dorsally.
- The penis is usually stubby and foreshortened with a dorsal curvature. It is more commonly associated with bladder exstrophy (exstrophy-epispadias complex) than in isolation and all are incontinent.
- In the isolated epispadias anomaly (primary epispadias), glanular or distal shaft types are usually continent, however, the rest (penopubic) have an associated bladder neck deficiency and are therefore incontinent.
- Other associations are extremely rare and do not require any further investigation.
- Issues encountered in adult life are retrograde ejaculation and difficulties with sexual intercourse.
- Reconstruction is carried out in infancy to keep in line with the normal milestones of achieving continence.
- Complications are similar to those seen in hypospadias and in addition, is a failure in achieving continence requiring redo surgery or alternate methods of achieving dryness. Other unsatisfactory outcomes are penile size and curvature.

Introduction

Primary epispadias is a rare condition and affects boys and girls with an incidence of about 1 in 120,000 with a male to female ratio of 5:1. In boys, it is characterized by the absence of dorsal foreskin, a wide urethral meatus, also situated on the dorsal aspect

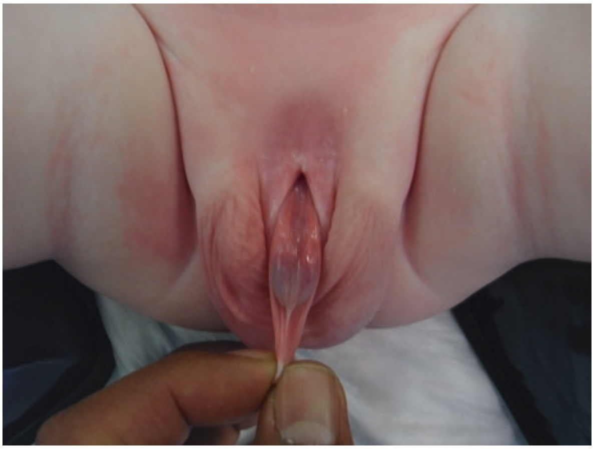


Fig. 5.6 Penopubic epispadias

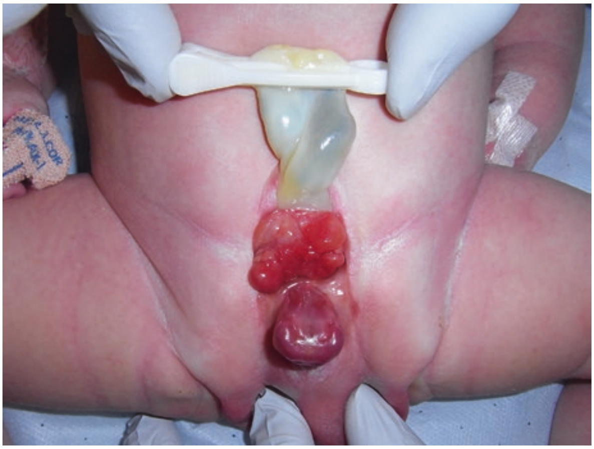


Fig. 5.7 Epispadias as part of the Bladder Exstrophy Epispadias complex

of the glans or shaft or root of the penis. The penis is wide, foreshortened and has a dorsal curvature. The glanular type can remain undetected until much later due to an intact foreskin but generally, the condition is fairly obvious. The more proximal types (pubic and penopubic) are associated with pubic diastasis and bladder neck deficiency, therefore, these children are incontinent (Fig. 5.6). Females usually present much later when investigated for primary incontinence. Most epispadias are in association with bladder exstrophy and hence termed exstrophy-epispadias complex with an incidence of about 1 in 50,000 (Fig. 5.7). Here the anterior abdominal wall and adjoining anterior bladder wall are absent with varying degrees of pubic diastasis.

Management

Children with primary epispadias or exstrophy-epispadias complex are otherwise healthy and do not have associated anomalies. Referral to a pediatric urologist is appropriate once the newborn has been stabilized within the first few days of life especially if it is an exstrophy-epispadias complex. Primary epispadias does not need urgent repair or intervention and can, therefore, be referred electively. Ultrasound of the urinary tract is the only investigation required, to rule out upper urinary tract anomalies although rare. Prophylactic antibiotics are usually commenced due to coexisting vesicoureteric reflux. Bladder outlet resistance is very poor in penopubic and exstrophy-epispadias complex hence continence surgery in early life is also to promote bladder growth and capacity which is an important contributor in achieving continence. In later life, a short and wide penis with curvature makes sexual intercourse difficult. An incompetent bladder neck promotes retrograde ejaculation and can cause fertility issues.

Surgical correction is usually a single stage procedure correcting the appearance, curvature and the bladder neck deficiency before 2 years of age to provide the opportunity with achieving normal milestones of continence.

Complications

These are similar to hypospadias and include urethro-cutaneous fistula, persistent dorsal penile curvature, ongoing urinary incontinence, and short penile shaft. Most of these are challenging except fistulas, which may be relatively easy to correct. Secondary procedures are usually employed to improve continence and may include endoscopic injection of bulking agents to the bladder neck, bladder neck reconstruction, slings and artificial sphincters all of which carry their own risks.

Concealed Penis

Key Points

- A variety of anomalies are included under this phenotype due to varying etiologies: (1). Congenital mega-prepuce (2). Buried penis due to excessive pre-pubic fat pad (3). Peno-scrotal transposition or webbing (4). Trapped penis within scarring following circumcision (5). Micro-penis (need Endocrine referral).
- Penile shaft is usually normal in size except those with true micro-penis. Circumcision is contraindicated in all the above and should be referred to a pediatric urologist.

Introduction

Concealed penis encompasses a variety of conditions including congenital megaprepuce, buried appearance due to an excessive pre-pubic fat pad, penoscrotal transposition and webbing, and iatrogenic following a circumcision (Figs. 5.8a, b and 5.9). In all the above the penile shaft itself is of normal size and is important to distinguish it from true micro-penis. The above conditions raise considerable anxiety in the minds of parents and an ill-advised circumcision becomes counterproductive. The repair or correction of these require the foreskin for redistribution and remodelling for adequate penile shaft cover. Most are diagnosed at birth and some become apparent later in childhood.

In the mega-prepuce variety, there is an excess of the inner prepuce and very little outer prepuce [3]. During the act of voiding urine collects within the inner prepuce giving the appearance of a volcano followed by overflow dribbling and is easily

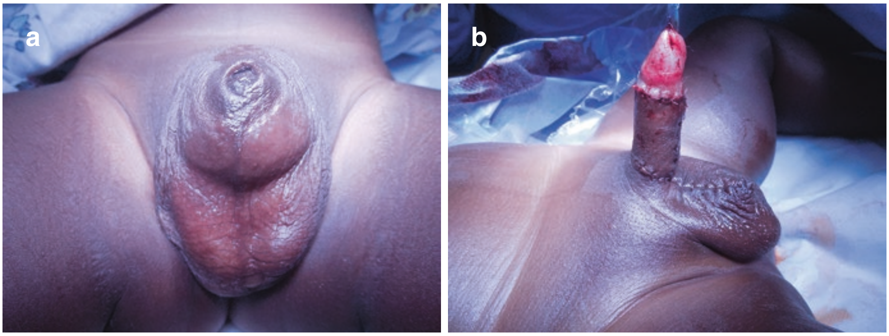
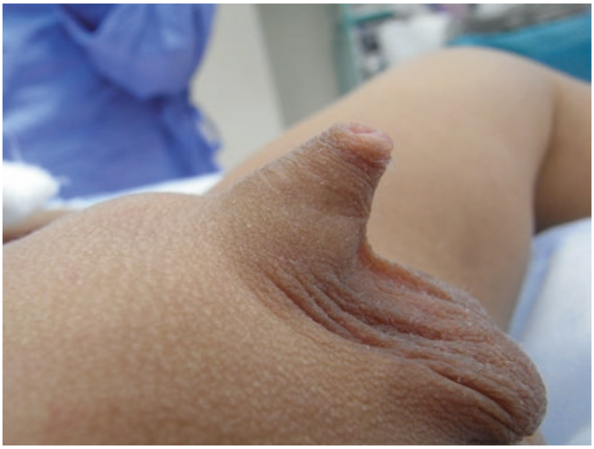


Fig. 5.8 (**a**) Congenital megaprepuce (CMP) creating a volcano effect and concealing the penile shaft on inspection. (**b**) CMP following correction

Fig. 5.9 Concealed penile shaft due to excessive prepubic fat pad



expressible. This collection under the skin is sometimes called a preputial bladder. They can at times present with recurrent urinary infections.

A true micro-penis is when the stretched penile length is less than two standard deviations (0.8 cm) below the normal (3.5 cm ± 0.4, Term in the USA). Endocrine input is necessary to evaluate for hypogonadism.

Management

The neonate is usually assessed by the pediatrician in the hospital at birth and any doubt in the external genital appearance should be followed by the advice to not circumcise the baby without being seen by the pediatric urologist. Significant concealed penis excluding micro-penis requires surgical reconstruction. A variety of procedures have been described for correction and are undertaken around a year of age. Mega-prepuce repair involves penile degloving, the release of the shaft skin and dartos or adventitia followed by redistribution of skin for shaft cover [4]. It is important to recognize that all of the outer prepuce and shaft skin should be preserved to achieve adequate and tensionless shaft cover. Excising the adventitia as much as possible from under the skin allows the skin to stretch. Fixing the skin cover to the base of the penis promotes better cosmetic appearance in the long term. Peno-scrotal webbing also needs the release of the shaft by the division of the web and scrotoplasty. Significant pre-pubic fat pad is generally best left alone but liposuction has been described with mixed results and is generally not recommended until after puberty with adequate motivation to lose and maintain appropriate weight to prevent recurrence. Scar entrapment of the penis following circumcision should be revised and can prove quite challenging due to shaft skin loss involved by scarring.

Complications

Recurrence of the concealed appearance seen with mega-prepuce repair may need redo surgery. This, however, should be attempted only after a period of initial expectant management as some may improve with time. The key element is recognition of the anomaly in the first instance and not attempting a circumcision as this would result in further loss of skin cover required to do a definitive repair, quite different from a standard circumcision. Redo surgery may require skin grafting or more complex skin flaps.

Benign Urethral Lesions in Boys

Urethral diverticulum or megalo-urethra, anterior urethral valves, syringocele, duplication, polyps, and congenital fistula are all rare.

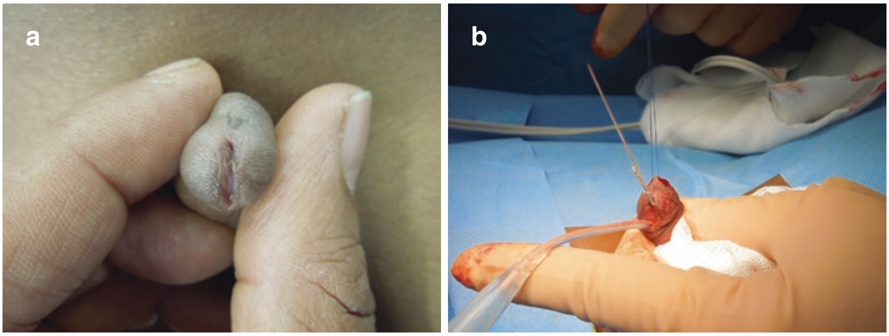


Fig. 5.10 (**a**) Duplicated urethra evidence by two openings on the glans. (**b**) Duplicated urethra with both openings cannulated

Meatal stenosis although uncommon, is more often seen in circumcised males and may be due to repeated abrasions of the exposed meatus while they were in nappies in early life. Presentation is in the form of a thin stream, straining, dribbling in older children, and at times dysuria and hematuria.

Urethral diverticulum, anterior urethral valves, and syringocele are considered a spectrum, and by some the same anomaly. Essentially it is caused by ectasia or cystic dilatation of the Cowper's gland ducts. These are bulbourethral glands that produce mucoid secretions homologous to the Bartholin's glands in females. Rupture of the cyst results in partial urethral obstruction and can present with a poor stream, dribbling, or urinary infection.

Anterior urethral diverticula are pouch-like saccular dilatations involving the floor of the urethra. Megalo-urethra is when it involves the entire urethra [5]. These may be associated with Prune belly syndrome and can produce abnormal voiding patterns and post void dribbling. They are diagnosed clinically due to a visible bulge of the penile shaft during the act of voiding and confirmed by a urethrogram.

Duplication anomalies of the urethra are rare and several types have been described including a dorsal moiety ending in a sinus, a 'Y' type duplication wherein the two moieties join somewhere proximally and the common channel entering the bladder, and finally a complete duplication of the urethra with sometimes a duplicated bladder as well (Fig. 5.10a, b). Two distinct streams may be noted and at times the ventral moiety may end at the anal verge or within the anal canal or rectum. Usually, the ventral moiety is dominant and more developed, while the dorsal is dysplastic or atretic.

Urethral polyps and fistula are extremely rare and the former present with obstructive symptoms, hematuria or infection. The latter are similar to those associated with hypospadias repairs.

Management

Most urethral lesions are symptomatic and fairly easy to diagnose clinically or with the help of a maturating cystourethrogram or by cystoscopy. Symptomatic ones are therefore best referred to a specialist for workup and treatment.

Polyps need excision with the base fulgurated by urethroscopy. Meatal stenosis is treated with a meatoplasty or meatotomy. Practices vary from an outpatient clinic procedure under local anesthetic to formal surgery under a general anesthetic. The choice of approach will depend on the extent of involvement, patient compliance, parent and surgeon preference. Meatal dilatations provide temporary relief and should generally be avoided if possible. Following correction, it is important to provide good analgesia. Meatal application of ointment for about 2 weeks with the help of a nozzle keeps the meatus open and prevents recurrence in the early days of wound healing. Follow up should be long term both clinically and with uroflow studies.

Fistulae are repaired no different from the ones seen following hypospadias repair. Diverticula, either posterior or anterior need complete evaluation and work up due to other associations. Syringocele is treated by endoscopic resection of the valvular leaflets however varying degrees of obstruction have been reported resulting in effects similar to posterior urethral valves such as bladder dysfunction and renal impairment. Saccular or other diverticula of the anterior urethra need excision and urethroplasty.

Y-type duplications can be quite challenging to treat as redo surgery is quite common [6]. The more simple ones such as the dorsal sinus can be well left alone and the common wall between moieties divided when the openings are quite close to each other.

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Chapter 6
Disorders of Male External Genitalia: Undescended Testis

M. V. Rodriguez, D. Kanabolo, and M. S. Gundeti

Introduction

Cryptorchidism, derived from the Greek words kryptos (hidden) and orchis (testicle) is the most common genital malformation identified at birth. Many scientific papers uniformly characterize undescended testes using the definition set by Scorer as testes less than 4 cm below pubic crest in full term males, and 2.5 cm in preterm males [1]. The testis may be located anywhere along the path of descent, these include intra-abdominal, inguinal (canalicular), scrotal or ectopic. The current standard of care, set forward by the American Urological Association is orchiopexy—surgical repositioning of the testis within the scrotal sac.

Epidemiology

Approximately 3% of term males and up to 30–45% of preterm males are affected by cryptorchidism at birth [2, 3]. This incidence declines during the first year in 75–90% of cases [4]. By 1 year of age, this figure approximates 0.8–1.2% [5–7].

It should be noted that environmental factors may play a role. A prospective study comparing cryptorchidism rates between Denmark and Finland found a 9.0% rate (95% CI 7.3–10.8) in Denmark and 2.4% (1.7–3.3) in Finland [8].

The true incidence of congenital cryptorchidism appears bimodal, tending to occur predominantly at birth and between ages 5–7. However, it is thought that these cases may be falsely diagnosed, due to a “retractile testes”. A retractile testis is commonly due to an overactive cremaster muscle. The cremaster reflex is strongest between ages 2 and 7, most commonly seen in boys between age 5–6 [9, 10].

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The testis may be manually replaced into the scrotal position and remain there temporarily. This feature differentiates retractile testis from the general phenomenon of ascending testes, which describes the change from previously intrascrotal testes to extrascrotal. This may be from multiple etiologies, including and not limited to: a hyperactive cremaster, inelastic spermatic cord, and fibrous processus vaginalis. The AUA recommends providers assess the position at least annually to ensure no secondary ascent [9].

Pathophysiology

Normal testicular descent happens by a complex sequence of events occurring in late pregnancy. The biphasic model describes the interplay of the caudal genitoinguinal ligament, also known as the gubernaculum, and the cranial suspensory ligament. The gubernacular bulb exhibits outgrowth and the gubernacular cord shortens. At 10 weeks' gestation, the gubernaculum deposits extracellular matrix at the caudal end of the gonad, forming a cone-like structure. The swelling anchors the developing testis to the inguinal region in fetal development. Concurrent androgen dependent regression of the cranial suspensory ligament occurs. At 26 weeks' gestation, the gubernaculum bulges through the inguinal canal, reaching the scrotum by 35 weeks. The caudal segment of the gubernaculum regresses to a scrotal ligament [11].

The etiology of cryptorchidism is less understood. Some suggest that a blunting of normal gonadotropin surge seen during gestation and in spontaneously descended testes may contribute to this condition [11, 12]. Others postulate that hypoandrogenism may be a cause [11]. The condition is associated with a 4% paternal and 6–10% fraternal concordance, suggesting a genetic component, though this may likely be polygenic and multifactorial [13].

Evidence for surgical management:

1. Men with an undescended testis appear to have lower sperm quality and counts when compared to males with bilateral descended testes [14].
2. The frequency of spontaneous descent is unclear. According to Thong et al., it is common by 1 year, approximately 76.5% in one series (91% premature n = 11, 69.6% term n = 23) published in 1998 [15]. A study published by Wenzler et al. in 2004 however, states that it is 6.9% at 1 year and beyond for patients initially presenting for evaluation of spontaneous descent before 6 months (n = 278) [16]. Mean testicular volume in undescended testes is impacted by 6 months [17]. This is theorized to be due to increased temperature (37 °C) and consequent loss of spermatogonial stem cells [16–18]. Thus, many surgeons suggest 6 months for definitive surgical repair.
3. Infertility rates worsen for bilateral undescended testes [17].
4. Improved physical examination of the testis allows earlier detection of malignancy [19]. Note: The contralateral descended testis appears to have no increased risk of malignancy [20].

5. The risk of testicular germ cell tumors, such as carcinoma in situ and testicular seminoma are increased 40×, compared to those with descended testis [21]. This may be due in part to multinucleated cells from impaired spermatogenesis [22]. Relative risk of testicular cancer in a cryptorchid testis repaired by age 12 is 2–3, but 2–6 when repaired after age 12. These rates appear to be proportionally higher in persistent intra-abdominal vs persistent inguinal testes (25%, n = 16 vs 7%, n = 44) according to one series [20].
    a. As such, orchiectomy should be considered in patients between 12–50 years of age, though this recommendation must be individualized to weigh risks and benefits [20].
6. A vast majority of undescended testes are associated with a patent processus vaginalis and thus increased likelihood of congenital inguinal hernia [23].
7. Torsion may occur relatively more frequently in the undescended testis during the first year of life. The mechanism for this is unclear, though two theories have been postulated. The first is thought to be due to spasms of the cremasteric muscles. The second due to abnormally larger size and distorsion of the normal dimensions, predisposing it to torsion [24].

Clinical and Surgical Management

Hormonal Therapy

Evidence has shown that the use of human chorionic gonadotropin (hCG) and luteinizing hormone-releasing hormone (LHRH) is not effective as a therapeutic measure to induce testicular descent. Its value might have been perceived when addressing low-riding testicles, a modality that gained popularity in Eastern countries at the time. Notwithstanding, the therapy was quickly abandoned given its discouraging results, poor success rates (8–60%) and numerous side effects (75%) [25].

In 2014 the American Urological Association released their last consensus for the management of undescended testis, stating that the use of hormonal therapy was not recommended to treat unilateral or bilateral cryptorchidism given the low response rates and lack of long-term efficacy [26].

Surgical Approach

Orchiopexy is the gold standard for the treatment of undescended testis. The advantage in treating these children include fertility preservation, testicular torsion/injury prevention, closure of the patent processus vaginalis, testicular cancer detection by making the testis more accessible during physical examination, as well as avoiding the psychological impact of having an empty scrotum [27, 28]. Imaging studies have no role in the diagnosis of cryptorchidism; even in nonpalpable testis due to its low specificity and sensitivity. On the contrary, laparoscopy is diagnostic and therapeutic, providing a definitive management for these patients.

If the testicle is palpable, the classic approach is through an inguinal or a scrotal incision based on the anatomic location of the testis. Both modalities allow for satisfactory access to low or high-riding testes. The decision to proceed with either approach is dictated by the surgeon's preference or training, as well as the anatomic location of the organ. The surgeon will also decide in agreement with the family if the procedure will be performed with either open or minimally invasive surgery [29].

In some circumstances, the testicle is too high that cannot be adequately descended, although there are several approaches that might help to bring it down. The most commonly used is the Prentis maneuver, which consists in mobilizing the spermatic cord medially by sliding the testis behind the spermatic vessels. This technique will provide an adequate cord length allowing better testicular mobilization to perform the orchiopexy, this is used as an adjunct to the open or laparoscopic approach.

Another procedure that can be useful in these cases is the Fowler-Stephens technique, with this approach, the internal spermatic vessels are divided laparoscopically with the purpose of mobilizing the testis as much as possible to achieve orchiopexy in either a one or a two-staged repair. The surgical principle is to preserve the collateral circulation (deferential/external spermatic vessels) which will eventually maintain the organ's blood supply [25].

In average, 25% of all cryptorquid testis are nonpalpable. Out of those, 50–60% will be intraabdominal, canalicular or peeping (close to the internal inguinal ring), 30% will be found atrophic or rudimentary in the inguino-scrotal area and the remaining 20% might be absent. Nonpalpable testes are diagnosed in the office, but a second evaluation needs to be performed in the operating room, the surgeon must examine the child under general anesthesia to confirm if it is palpable or not.

Diagnostic laparoscopy is considered the most accurate method of surgical exploration in these cases since it provides a reliable way to evaluate the location of the testis, whether an inguinal hernia is associated or not, and the external appearance of the testicular vessels (blind-ending vessels might be an indicator of an absent testis) [26, 30].

Sporadically, intraabdominal nonpalpable testes are not able to reach the scrotum. Therefore, the solution is to divide the testicular vessels, wait for 6 months until the collateral blood supply has had time to develop, and perfom a second stage surgery in 4–6 months—Fowler–Stephens staged orchiopexy. By this time, sufficient circulation will allow adequate testicular mobilization and successful orchiopexy [26].

In regards to testicular nubbins, there is still controversy in the surgical management of these children, some providers will choose to perform orchiectomy of the

Table 6.1 Classifications of cryptorchidism

Palpable [2, 3, 32–37]		
Type	Description	Frequency in male pediatric population
Undescended cryptorchidism	Congenital failure of testicular descent	3–45%
Retractile testis	Form of acquired undescended testis whereby the testis may be manually replaced into the scrotal position and remain there temporarily. Due to hyperactive cremaster muscle	3.9% (males between ages 7 and 12)
Ascending testis	Child with low undescended testes experiences testicular ascent from previously intrascrotal position to extrascrotal. Commonly detected in periods associated with increased linear growth	2.2% (males between ages 6 and 13)
Iatrogenic cryptorchidism	Suprascrotal testes after inguinal hernia repair	0.8–2.8% (as percentage of those undergoing inguinal hernia repair)
Non-palpable testes [38–42]		
Location	Description	Frequency as a percentage of non-palpable testes
Abdominal/ transinguinal	Undescended testes hidden within peritoneal space or inguinal canal	25–50%
Atrophic "vanishing"	Regression and disappearance of the testes early in pregnancy	15–40%
Extra-abdominal	Limitation by body habitus, patient cooperation, testes size	10–30%

cryptorchid testis, while others will rather to leave them in situ [26, 31]. Our institutional preference is to explore, remove the nubbin, and fix the contralateral testis to prevent the possibility of torsion and loss of the solitary testis (Table 6.1).

Indications for Referral

- If a child is diagnosed with cryptorchidism, and testicular descent has failed to occur by age 6 months (corrected by gestational age), an appropriate referral should be made to the pediatric urologist. Imaging studies are not necessary, given that these studies seldom assist in the definitive management of these children [25, 26].
- If a boy is diagnosed with a cryptorchid testis after 6 months of age (corrected for gestational age), an appropriate consultation should be made to address the possible diagnosis of acquired undescended testis [26].
- For all phenotypic male newborns diagnosed with bilateral nonpalpable testes, a multidisciplinary workup should be prompted to evaluate disorders of sex devel-

opment. Concomitant severe hypospadias should also raise suspicion for disorders of this kind [26, 43].

- Absence of Müllerian Inhibiting Substance (MIS) in addition to elevated LH/FSH hormones in a patient with bilateral nonpalpable testes strongly suggest testicular agenesis. Congenital adrenal hyperplasia must be ruled out first [26, 44].

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Chapter 7

Pediatric Surgery and Urology for Primary Care Physicians: *Circumcision*

Duncan T. Wilcox

Key Points

- A non retractile foreskin is common at birth and majority of boys will have a fully and easily retractable foreskin by physical maturity.
- Circumcised males are at lower risk of UTI's, HPV 2 and penile cancer. However the number required to treat to prevent 1 UTI in the first year of life is significant.
- Absolute indications for circumcision is balanitis xerotica obliterans, Paraphimosis refractory to manual reduction and Recurrent balanoposthitis with failed conservative management.

Natural History of the Prepuce

The prepuce is apparent prenatally at 8 weeks of gestation as a ridge of thickened epithelium. The prepuce grows forward over the glans to completion by 16 weeks of gestation. Early in gestation there is no separation between the epithelium lining the prepuce and glans. Preputial adhesions are a characteristic of normal development with initiation of spontaneous separation by desquamation late in gestation. The rate of preputial separation after birth is variable with up to 70% of boys having some degree of preputial adherence at 5 years of age. Consequently a "non-retractile foreskin" or more appropriately described "physiologic phimosis" at birth is normal (Fig. 7.1). The majority of boys will have a fully and easily retractable foreskin by physical maturity.

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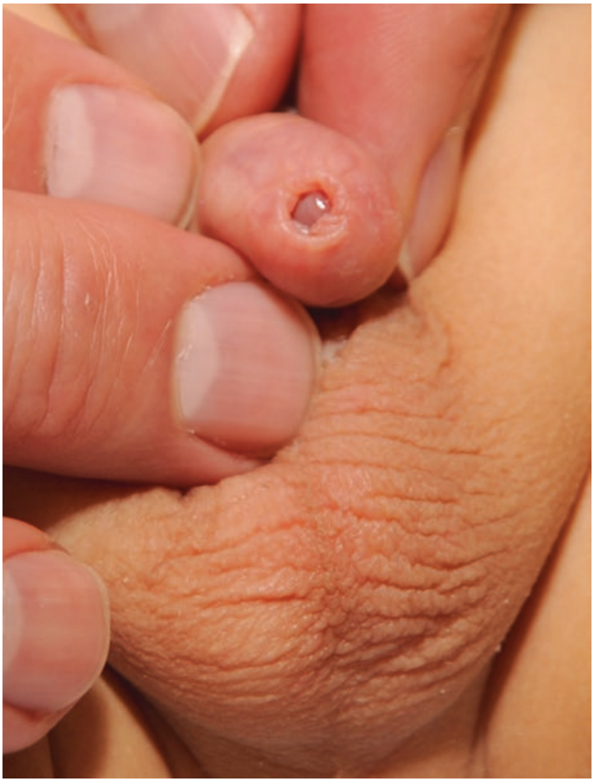

Fig. 7.1 Normal appearing foreskin

Benefits of Circumcision

1. UTI's: Circumcised males infants are at a lower risk of UTI's than uncircumcised infants. The effect of circumcision on UTI's has been studied primarily in infants because they have a higher prevalance of UTI's than older males. The incidence of UTI in circumcised male infants has been reported at 0.10% versus 1% in uncircumcised male infants. Despite the decreased risk of UTI's in circumcised infants it has been reported that as many as 195 circumcisions would be needed to prevent one hospital admission for UTI in the first year of life.
2. Cancer: Neonatal circumcision has a protective effect against invasive penile cancer.
3. STD's: Circumcision has been demonstrated to reduce the incidence of HSV-2 infection and the prevalence of HPV infections. Circumcision is also associated with a reduced risk of HIV infection. Although practicing safe sex is the most reliable and recommended method of preventing STD's

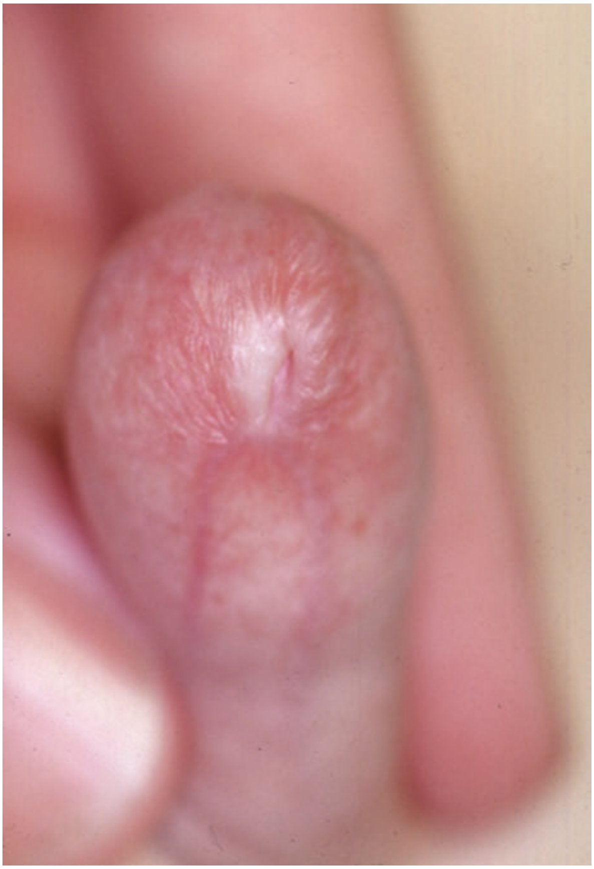

Fig. 7.2 Scarred foreskin seen in patients with BXO

Absolute Indications for Circumcision

1. Paraphimosis refractory to manual reduction
2. Recurrent balanoposthitis with failed conservative management
3. Balanitis xerotica obliterans (Fig. 7.2)

Relative Indications for Circumcision

1. Persistent phimosis
2. To reduce the rate of STD's and HIV particularly in sub Saharan Africa
3. Cultural and Religious preference

Surgical Options

Circumcision is most often performed on neonates using the Gomco clamp or one of the plastic disposable devices such as the Hollister Plastibell. Circumcision should be performed on an infant no earlier than 12–24 h old. Various options are available for analgesia ranging from allowing the infant to suck on a sucrose solution, topical anesthetics (e.g. EMLA cream), to various local blocks (e.g. dorsal penile nerve block or ring block) using 1% local anesthetic without epinephrine. It is important to free the glans completely from the inner mucosal layer of the foreskin. It is essential to observe that an equal amount of foreskin is present circumferentially when pulling the foreskin into the clamp. Care should also be taken to pull a proper amount of foreskin into the clamp to avoid taking too much or too little foreskin with the circumcision. Routine post-circumcision care includes application of an ointment (e.g. triple antibiotic) to the wound for several days and resumption of normal bathing.

After several months of age circumcision should be performed as a formal procedure with general anesthesia in addition to a local anesthetic. Several different surgical techniques are employed and are a matter of individual preference. Circumcision clamps are not recommended in the older patient as the vessels become larger and are not easily sealed by compression. As in the neonate, care is taken to separate the prepuce from the underlying glans to prevent formation of persistent skin bridges between the glans and penile skin. The outer and inner layers of preputial skin are excised separately watching closely to excise sufficient but not too much preputial skin. During the procedure meticulous haemostasis is maintained (e.g. bipolar diathermy). Several methods of skin closure are described including the use of rapidly absorbable sutures to avoid suture tracts, subcuticular sutures, as well as the use of tissue glues.

Contraindications to Circumcision

Patients with hypospadias, epispadias, or ambiguous genitalia should not undergo neonatal circumcision as their foreskin may be required for reconstructive purposes. Circumcision should also be avoided in those with chordee without hypospadias, hidden penis, webbed penis, micropenis, dorsal hood deformity, and megaprepuce. If an anomaly is detected after the dorsal slit is made it may be better to stop the procedure rather than proceeding with the circumcision, however if the anomaly is minor circumcision should proceed as the foreskin is very rarely needed if further reconstruction is necessary. One should also inquire about a family history of bleeding disorders prior to proceeding with circumcision to avoid excessive postoperative hemorrhage.

Complications of Circumcision

1. Bleeding: usually from the frenulum or from a vessel along the penile shaft which is generally controlled with compression.
2. Wound infection

3. Penile adhesions
4. Meatal stenosis: most common late complication.
5. Too much penile skin removed: if this occurs, apply triple antibiotic ointment to the open wound. Usually most of the skin will grow back bridging the defect and rarely is immediate skin grafting necessary.
6. Too much penile skin left: revision circumcision is often requested by parents for an incomplete circumcision.
7. Urethral injury
8. Partial removal of glans: has been reported with the Mogen clamp and should be immediately sutured back in place
9. Urethrocutaneous fistula: low incidence, thought to be secondary to poorly placed suture at frenulum in an attempt to control bleeding.
10. Penile necrosis: rare, can result from thermal injury secondary to cautery.

Conclusion

Circumcision is the most common operation performed world wide. In a few boys' circumcision is clinically necessary but in the vast majority of boys circumcision is performed for cultural or religious reasons. However, circumcision should not be performed without the families understanding that there is a significant complication rate and reoperation rate.

Suggested Reading

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Chapter 8
External Genitourinary Concerns in Females

Elizabeth B. Yerkes

Key Points

Interlabial masses:

- Interlabial masses may be associated with the urethra or vagina and may be congenital or acquired.
- The appearance can be alarming to parent and provider, but lesions are most commonly benign and easily managed.
- A diagnosis can generally be discerned based upon anatomical landmarks, clinical presentation and tissue characteristics. Imaging is not generally required.

Labial adhesions:

- Secondary agglutination of the relatively fragile labial and vestibule mucosa obscures the otherwise normal urethral and vaginal orifices.
- Medical or surgical management not generally needed due to spontaneous separation with puberty. Indications for management include:
  - Urinary retention (very rare)
  - Urinary tract infection or persistent UTI symptoms
  - Refractory post-void dribbling in toilet trained child
- Benign adhesions are important to distinguish from congenital labial fusion or urogenital sinus, as the latter will not resolve with conservative or expectant management and require more complex surgical management.

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Evaluation of Interlabial Masses

Interlabial masses, such as urethral prolapse, paraurethral cyst, prolapsed ureterocele and sarcoma botryoides, can generally be diagnosed without expense of imaging studies. Diagnosis is based upon the age and presenting symptoms or historical elements, relationship of the mass to surrounding anatomical landmarks, and tissue color and characteristics.

Imaging with ultrasound may be considered in the following scenarios:

- If relevant to further evaluation and management (ureterocele)
- If diagnosis or need for management is not clear
- If lingering concern for a serious diagnosis (sarcoma botryoides)
- In follow-up of initial urinary tract dilation, if identified (ureterocele, hydrocolpos)

Urethral Prolapse

Define (What/Who)

Circumferential prolapse of urethral mucosa, as if a sock is cuffed inside out. Classic presentation is in 4–5 year old girl, often but not exclusively African American. History of acute hard cough or constipation is generally present. Affected girls note pressure or fullness in the vulvar area and tenderness with wiping. Blood spotting in underwear occurs spontaneously or with wiping.

Distinguish (Where/What)

- Prolapsed tissue in donut configuration in expected location of urethral meatus (Fig. 8.1)
- Varying degrees of congestion and ischemia so may appear beefy red, purplish or speckled with superficial necrosis.
- Radial vasculature pattern of exposed tissue
- Imaging not indicated

Manage (Why/How)

Conservative management and topical preparations are generally successful in encouraging retreat of the prolapsed tissue.

- Tub soaks
- Address stool retention or underlying respiratory factors
- Topical treatment with bland ointment to restore moisture and protect the mucosa.
- Topical estrogen cream to the prolapsed tissue BID for up to 2 weeks is also a common intervention.

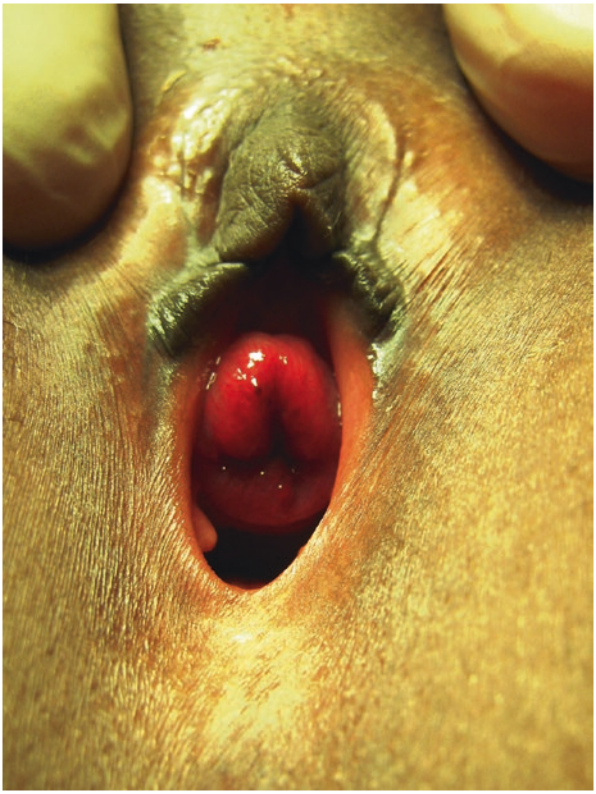

Fig. 8.1 Urethral prolapse: circumferential prolapse and edema of urethral mucosa. Note the donut shape with central urethral meatus. Also note patent hymen

For ongoing pain and congestion refractory to conservative management or for severe ischemia/necrosis, surgical management is undertaken.

- Prolapsed tissue is excised in quadrants
- Vestibular mucosa approximated to urethral mucosa with resorbable sutures.
- Manage underlying condition (stool retention)

In some cases, the prolapsed mucosa will regress but leave a small lip of residual prolapse at 6:00. In these cases, continue to manage stool retention and treat mild irritative symptoms with bland ointment. Ultimate regression or maturation of tissue is anticipated.

Paraurethral Cyst (Skene's Gland Cyst)

Define (What/Who)

Paraurethral cyst is an obstructed Skene's gland discovered in the neonatal period. Maternal hormones in pregnancy stimulate mucus production, resulting in the milky or whitish-yellow fluid in the cyst.

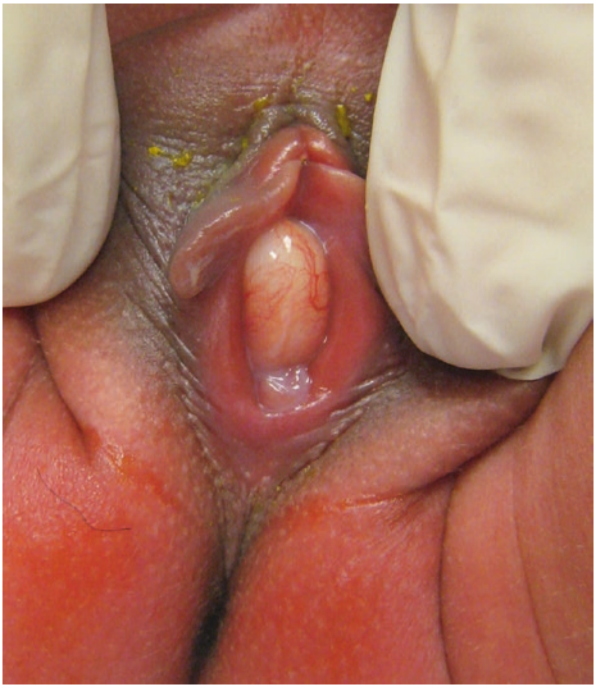

Fig. 8.2 Paraurethral cyst: Whitish-yellow parameatal cyst with superficial vessels. Meatus is not visible. Note hymen tissue, estrogenized by maternal hormones, immediately adjacent

Distinguish (Where/What)

- Round smooth thin walled cystic structure with overlying small blood vessels, eccentric (ventrolateral) to urethral meatus (Fig. 8.2)
- Milky or whitish-yellow fluid
- Vaginal orifice and hymen may be obscured by large lesions but not obstructed or involved, distinguishing it from the whitish bulge with smooth membrane that characterizes neonatal imperforate hymen
- Imaging not indicated.

Manage (Why/How)

Reassurance is the primary intervention. Typically no active intervention is required, as spontaneous rupture and resolution generally occurs in the first several weeks. If it persists, a nick with small needle can drain the fluid without recurrence. Occasionally older girls are noted to have a cup-like pocket at the urethral meatus—this may represent a previously decompressed cyst.

Ureterocele Prolapse

Define (What/Who)

Ureterocele is a congenital cystic dilation of the distal ureter associated with anomalous development at the ureteral orifice. These are most commonly diagnosed prenatally and majority are treated postnatally to relieve the obstruction. Most are associated with the upper pole of a duplicated collecting system, and co-existent vesicoureteral reflux to other systems may be present.

A large ureterocele can prolapse down the urethra during voiding, resulting in congestion of the ureterocele wall, but more importantly, bladder outlet obstruction.

Distinguish (Where/What)

- Prolapsed tissue comes per urethra and assumes an eccentric position relative to the urethral meatus
- Large masses fill the vestibule, obscuring visualization of the hymen but otherwise not involving vagina
- Exposed tissue is bladder mucosa, which becomes congested and edematous and less moist
- Progression from normal mucosa to deep red to purplish hue (Fig. 8.3)
- Imaging not specifically needed to support the suspicion by exam but prudent prior to intervention if ureterocele not previously known

Manage (Why/How)

Prolapse may be fleeting, but if congestion occurs, the prolapse will need reduction. Manual reduction is not likely to be successful as tissue changes progress. If a diagnosis of ureterocele is previously known from prenatal testing or from urinary tract infection, bedside incision is an option. However, incision in the operating room is standard intervention. With incision, the ureterocele decompresses and retreats. Elective endoscopic incision is recommended if the prolapse spontaneously reduces. Follow-up imaging with ultrasound and possibly VCUG and/or nuclear renal scan will be undertaken. Depending upon the overall anatomy and function, more definitive open reconstructive procedures (ureterocele excision and ureteral reimplantation or heminephrectomy) may be required in the future.

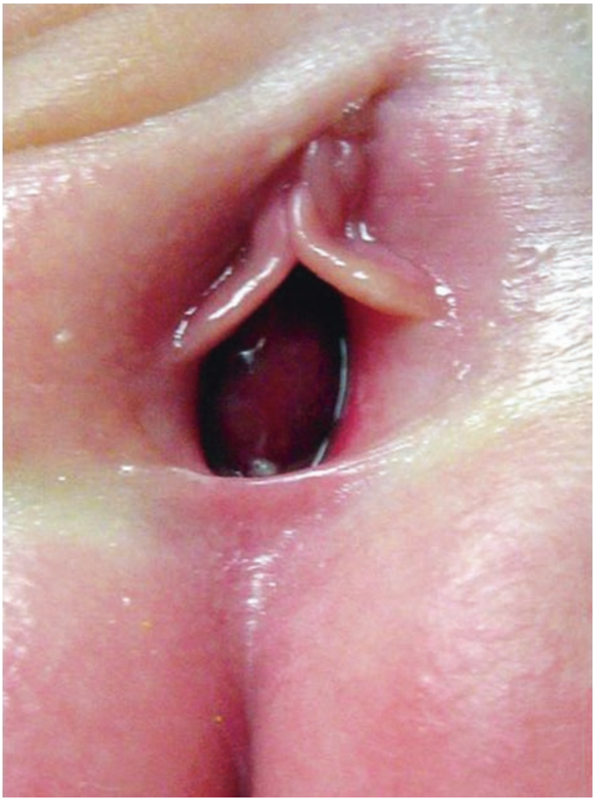

Fig. 8.3 Ureterocele prolapse: characteristic anterior interlabial mass with purplish discoloration consistent with congestion. Tissue emanates from meatus and sits eccentric to meatus

Sarcoma Botryoides (Rhabdomyosarcoma)

Define (What/Who)

Embryonal rhabdomyosarcoma originating in the vaginal wall occurs most commonly prior to 2 years of age but nearly uniformly before 5 years. After sufficient growth to present itself as an interlabial mass, that protrudes through the hymen as a cluster of nodules resembling a bunch of grapes. Bleeding or passage of necrotic tissue may lead to examination under anesthesia and earlier diagnosis. Urinary retention or voiding difficulties may also occur with a large mass due to pressure on the bladder outlet.

Distinguish (Where/What)

- Unique multinodular appearance as a bunch of grapes (Fig. 8.4)
- Bleeding and superficial necrosis of exposed tissue
- Urethral meatus in normal position but less easily seen with a large mass
- Hymen normal but obscured

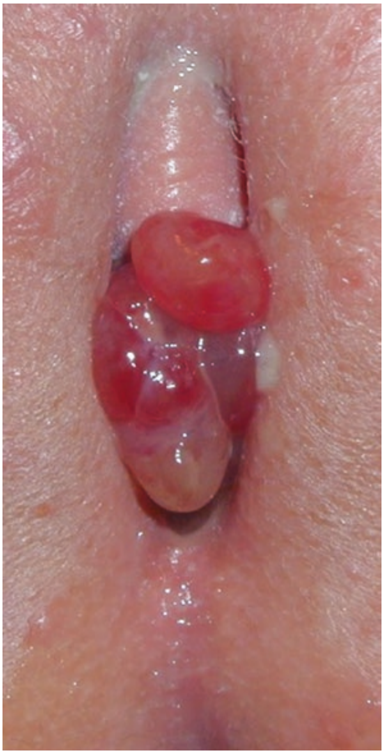

Fig. 8.4 Rhabdomyosarcoma (Sarcoma botryoides): interlabial malignancy with multinodular appearance reminiscent of a bunch of grapes

- Imaging may have occurred in course of investigation of symptoms but appearance generally prompts diagnosis
- 3-dimensional imaging indicated for staging after diagnosis made

Manage (Why/How)

Upon suspicion for vaginal lesions, the child should be urgently referred to urology. Biopsy, along with cystoscopy and vaginoscopy, is performed to make a histologic diagnosis and to ascertain primary organ of origin (vagina or bladder). Other rare vaginal malignancies may present similarly. Other than for very localized tumors, pediatric oncologic protocols generally involve chemotherapy and radiation with surgical management reserved for refractory disease.

Imperforate Hymen

Define (What/Who)

Imperforate hymen is the failure of the hymen to cannulate embryologically, resulting in trapped mucus or menses.

The obstruction may be apparent during neonatal period due to maternal estrogen stimulation (mucocolpos) or after menarche (hematocolpos). The vagina may be dilated enough to create a palpable abdominal mass and hydronephrosis related to compression of the ureters.

Distinguish (Where/What)

- Dome-like bulging, partially ballotable mass filling the vestibule (bulging nature distinguishes from vaginal agenesis) (Fig. 8.5)
- Urethral meatus normal position and unaffected
- Labial complex typical in appearance

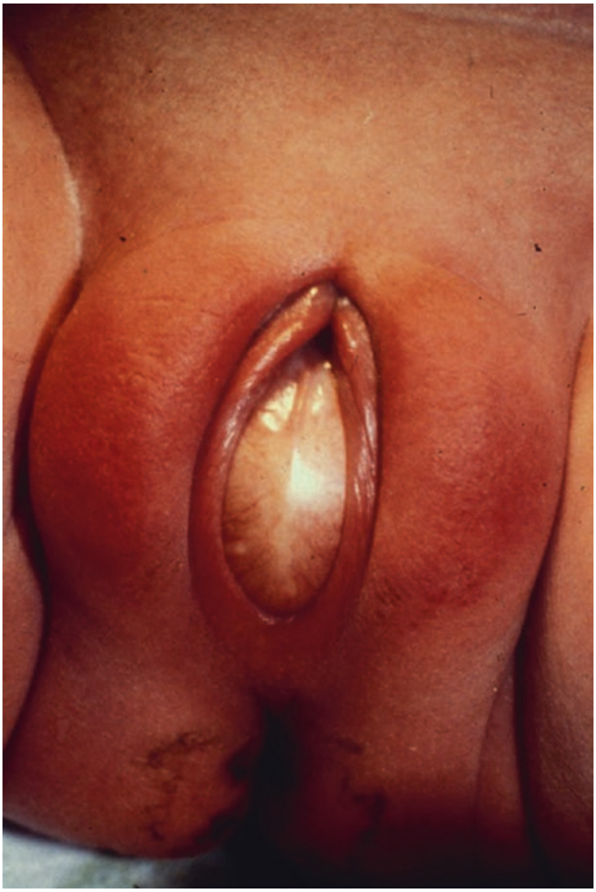

Fig. 8.5 Imperforate hymen in neonate: whitish mucus related to maternal hormones distend the hymen. Similar in character to paraurethral cyst but different shape. Rim of hymen not present and urethral meatus is noted anterior to obstruction

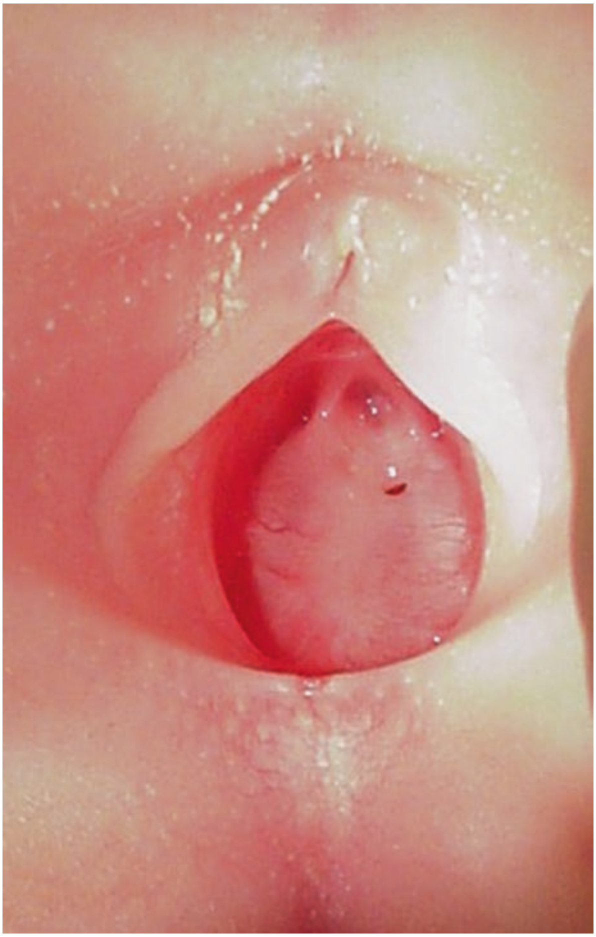

Fig. 8.6 Microperforate hymen: Note the urethral meatus and the small left anterior hole in the hymen. Membrane is pearly and somewhat translucent prior to estrogenization at puberty

- Rim of hymen not visible at margin of mass, distinguishing imperforate hymen from an intermittently bulging mass in the distal canal (very low transverse septum versus Gartner's duct cyst)
- Microperforate variants also occur and do not cause substantial obstruction but may be picked up as a pearly, mobile membrane prior to puberty (Fig. 8.6) and thick estrogenized membrane after puberty.

Manage (Why/How)

Once other introital masses have been ruled out, the hymen is incised widely longitudinally (infancy) or in cruciate fashion (post-puberty) to release the fluid. Neither dilations nor further interventions are required.

Gartner's Duct Cyst

Define (What/Who)

Cystic dilation of Wolffian duct remnant in the anterolateral vaginal wall can be associated with ectopic insertion of ureter of a single system or congenitally duplicated system. Most common presentation in this case would be continuous urinary incontinence unrelated to voiding, as the cyst spontaneously ruptures during development. Diagnosis may also be suspected but not confirmed during evaluation of antenatal urinary tract dilation. An uncommon presentation is a neonatal interlabial mass that protrudes intermittently or that is more evident with crying or straining.

Distinguish (Where/What)

- Small round mass intermittently presents at opening of vaginal canal
- Hymen normal
- Vaginal mucosa colored or slightly bluish related to the fluid density
- Imaging (ultrasound) of the upper and lower urinary tract and perineum help clarify the diagnosis
- Imaging with MRI is likely definitive but the risk of anesthesia for imaging is balanced against whether intervention is planned

Manage (Why/How)

Expectant management in an infant is very reasonable, as spontaneous involution may occur. Local surgical management without clear symptoms could lead to additional procedures later if incontinence occurs. If the cyst were related to a functional upper pole moiety, surgical management via ureteroureterostomy or extravesical ureteral reimplantation will help preserve the renal function and address the anatomical concern.

Labial Adhesions or Labial Agglutination

Define (What/Who)

The labia minora and inner rim of the labial complex are secondarily agglutinated, as opposed to congenitally fused. The adhesions generally do not form until the tissue benefits of maternal estrogen have faded. Thereafter, the moist and irritating diaper environment likely impacts the integrity of the less robust labial mucosa and adhesions may develop between the anatomically closely approximated structures.

The extent of agglutination varies, but adhesions may be present all the way to the posterior fourchette and with only a small aperture at the base of the clitoris. The adhesions form a wall or curtain that obscures inspection of the urethra and hymen and therefore also results in momentary filling of the vestibule with urine during voiding.

This temporary tissue distortion and chemical irritation results in UTI-type symptoms. True UTI is difficult to diagnose due to difficulty with clean specimen collection. Likewise, post-void dribbling of residual vestibular urine may be noted in toilet trained girls. Interestingly, deviation or spraying of the urinary stream is not a common complaint.

Distinguish (Where/What)

- Most commonly a narrow gray line between the labial minora (Fig. 8.7)
- Labial skin rather than labial or vestibular mucosa visible
- Although labial adhesions can be more dense, most are easily distinguished from true labial fusion and urogenital sinus by the character of the midline skin (Fig. 8.8)
- Thick adhesions, particularly if neovascularity noted crossing the midline, raise suspicion for friction or repeated separation associated with sexual abuse

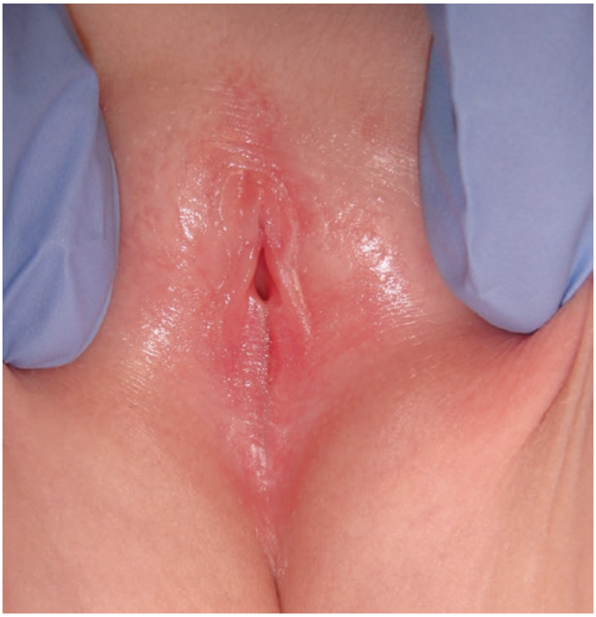

Fig. 8.7 Labial adhesions: labial and vestibular tissue agglutinated with anterior orifice below clitoris. Note the line of adhesion. Adhesions are incomplete anteriorly—labia minora would be pulled more to midline as adhesions progress in depth

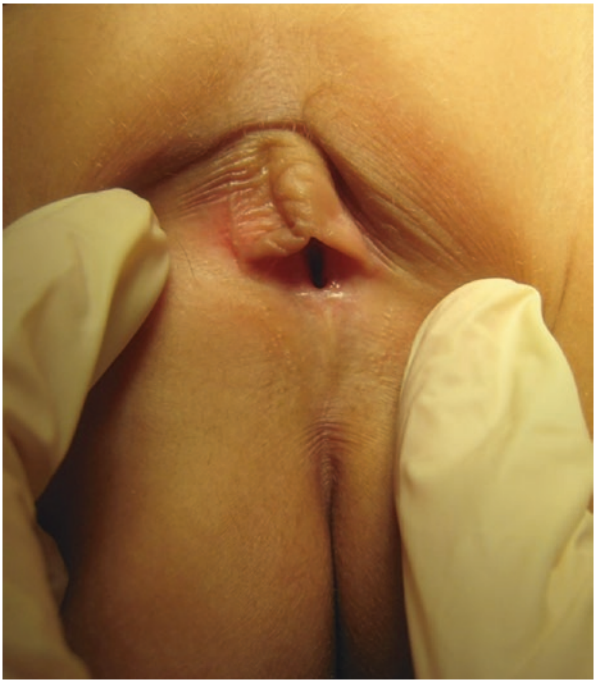

Fig. 8.8 Labial fusion as form of persistent urogenital sinus: Midline tissue is thick and does not have mucosal characteristics. There is no midline depression

Manage (Why/How)

Expectant management with perineal care and use of bland ointment will minimize progression and assist with tissue health but will not necessarily encourage resolution. Too much attention to the genital area (for the sake of the adhesions), resulting in anxiety and fearfulness on the part of the child, is counterproductive and medically unnecessary for most cases. This recommendation should be individualized. Avoidance of constipation is also likely beneficial for minimizing vulvar irritation.

Medical or surgical intervention is not typically instituted due to spontaneous lysis with return of estrogen at puberty. Indications for management include:

- Urinary retention (very rare)
- Urinary tract infection or persistent UTI symptoms
- Refractory post-void dribbling in toilet trained child

Topical treatment options are estrogen cream or high potency glucocorticoid ointment (such as 0.05% betamethasone dipropionate). Each should be used with caution, in a thin layer with gentle pressure directly to the adhesion rather than indiscriminately on the labia majora. Application is twice daily for 2 weeks and no more than one month. Detailed instructions to the parent will minimize risk of secondary local tissue changes and pigmentation or breast changes related to estrogen, as well as systemic absorption and influence on the Hypothalamic-Pituitary axis with glucocorticoids.

Manual lysis of the adhesions can proceed after failed medical therapy or by parental preference. This can be accomplished in the office with topical anesthetic for thin adhesions or can be addressed quickly in the operating room with less memory of the event. The latter also had the advantage of placing a few sutures to reapproximate the epithelium.

Regardless of the form of intervention, application of bland ointment for BID for 3 months thereafter is prudent to reduce recurrence.

Enlarged Clitoris

Enlargement of the clitoris may be present at birth, and in that case requires multidisciplinary evaluation for congenital adrenal hyperplasia or a difference of sex development (DSD)/ intersex. Newborns and premature babies in particular may have a prominent clitoral hood and labia minora that can be confused with clitoromegaly. If there is any question, a limited endocrinologic investigation, in consultation with an endocrinologist, may be indicated to rule out a life-threatening adrenal condition or other androgen producing state.

With secondary enlargement of the clitoris in infancy or childhood, evaluation by endocrinology and urology with adrenal and androgen labs as well as assessment for advanced bone age is indicated. A karyotype may also be recommended. A large percentage of girls and women have prominent labia minora, so assessment of the glans size and true clitoral length is helpful.

Infiltrative conditions, such as neurofibromatosis, or exogenous exposure to androgens also cause clitoral enlargement.

Suggested Reading

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Chapter 9

Disorders of Continence: Lower Urinary Tract Dysfunction

Nathalie Webb, Janet Chase, Marjorie Burgess, and Felix Grusche

Key Points

- Up to 10% of school age children have lower urinary tract symptoms (LUTS)
- Common conditions resulting in LUTS are overactive bladder (OAB), underactive bladder (UAB), voiding postponement (VP) and dysfunctional voiding (DV)
- Evaluation of LUTS should include a comprehensive history, physical examination and assessment of micturition
- Treatment is multimodal and multidisciplinary and must be tailored to the needs of the individual child

Introduction

Lower urinary tract (LUT) disorders are common in childhood, with up to 10% of school aged children presenting with issues of incontinence, recurrent urinary tract infection or both. Investigation may reveal underlying structural or pathologic causes in some, but for most the problem is functional. LUT dysfunction causes both physical and psychological morbidity to an increasing extent the longer it persists. Early and effective management is needed to mitigate longterm adverse impacts.

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LUT dysfunction is managed by diverse range of health care disciplines and specialties. The International Children's Continence Society draws together these multidisciplinary perspectives and provides a shared language of standardized terminology to reduce semantic confusion and allow meaningful comparison of outcomes from different interventions [1]. This chapter will employ the standard ICCS descriptive framework for LUT dysfunction; considering clinical entities of overactive bladder (OAB), underactive bladder (UAB), dysfunctional voiding (DV) and voiding postponement [2].

Lower Urinary Tract Symptom (LUTS) Terminology

Incontinence means involuntary leakage of urine. It may be continuous or intermittent and occur by day (daytime incontinence) and/or by night (enuresis). It is considered abnormal and significant in children 5 years or older, wet at least once per month for duration of 3 months or longer [2].

Frequency relates to how often the child voids. It is variable and influenced by age, urine volume and bladder storage ability [3]; with 3-8 voids per day accepted as normal [4–6].

Urgency refers to a sudden and unexpected, immediate and compelling need to void [6]; a common symptoms of overactive bladder.

Nocturia is waking at night to void. This is common among school-aged children [5] and not necessarily an indication of LUT dysfunction.

Enuresis refers to intermittent incontinence which occurs exclusively during sleeping periods [1]. Children who have intermittent incontinence while both awake and asleep have both daytime incontinence and enuresis. Enuresis is further explored in Chap. 12.

Lower Urinary Tract Dysfunction

Achievement of bowel and bladder control is an important developmental milestone in children, usually achieved between 3½ and 5 years. Girls generally develop control earlier than boys and daytime control usually preceeds nighttime [7]. Problems with this phase of normal development manifest as a heterogenous array of storage and voiding symptoms, grouped together under the umbrella term of 'lower urinary tract dysfunction' or LUTD. Longitudinal population studies show that the prevalence of LUT dysfunction decreases with age, indicating some natural resolution. However the impact of incontinence on the child and their family cannot be overstated.

Various studies have found 7–10% of school aged children present for medical attention due to urinary incontinence and/or recurrent urinary tract infection [8–10]. True prevalence is estimated to be higher, with many children suffering in silence

[11, 12]. There is a tendency toward increasing prevalence in more recent times, possibly illustrative of raised awareness and greater access to help [7]. Some of these children will be found to have underlying structural or pathologic causes, but most have a functional disorder of the lower urinary tract. Children with neuropsychiatric conditions such as attention deficit hyperactivity disorder (ADHD), have higher rates of functional incontinence.

In addition to the impact of chronic disease with extended need for medical interaction, these children have symptoms which are publically evident and for which general society is often intolerant. Resultant significant psychosocial comorbidity is well documented [13–17], compelling proactive and effective intervention. While there is a rate of spontaneous improvement with time [18], subgroups of persistent and relapsing symptomatology have a less predictable path [7]. As is currently not possible to accurately identify the likelihood of spontaneous improvement in any given child, 'just waiting' is not an option.

Evaluation of LUTS in Children

Children with LUT symptoms usually present in the non-acute, primary care setting. Assessment should involve a detailed structured history and a bladder diary. Physical examination should focus on urologic, neurologic and genital systems. Initial investigations include urinalysis and urinary tract ultrasound with measurement of pre and post-void bladder volumes [19]. If available, uroflowmetry can be a valuable addition. This screening should identify patients with neurologic or structural anomalies of the urinary tract, who require referral for specialised care.

History

General medical history should include antenatal and birth history, as well as review of age-appropriate developmental milestones and comorbidites. History of urinary tract infections is important, as are indicators of bowel function. A pictorial Bristol stool scale can be a useful tool for evaluating bowel function in children.

Exploration of the child's continence history needs to involve clear distinction between intermittent and continuous wetting, as well as between wetting while awake or asleep and before or after voiding. History of toilet-training attempts and ages can be useful to distinguish children at risk for LUT dysfunction. Quantification of urine loss volume is subjective and not especially reliable [19].

Questionnaires and scoring systems are used far less frequently in assessment of incontinence in children than they are in adults, with experienced clinicians relying on individualised history taking. However, there are validated instruments available that are useful both as an adjunct in diagnosis and a quantitative measure of change in evaluating management [20–23].

EBC = 30 x (age in yrs + 1) ml

Fig. 9.1 Simple formula for expected bladder capacity for age (EBC) [24]

Bladder Diary

Oral fluid intake, voiding frequency, voided volumes, wetting frequency, urge and reactions to urge are vital pieces of evaluative information, but recall alone is unreliable. A bladder diary is a simple and non-invasive method of objectively assessing these aspects, with the child or parent recording fluid intake (nature, volume and time), voided urinary volumes, episodes of urgency and episodes of wetting (large or small). Voided volumes must be compared with expected bladder capacity *for age* (Fig. 9.1), to adequately characterise the child's voiding history.

Two days' record is usually sufficient [25], although a lengthier diary without volume measurements can provide broader information: documenting frequency of nocturia, daytime incontinence, nocturnal enuresis, other LUT symptoms and bowel pattern: frequency, consistency (Bristol scale) and encopresis [1]. In addition to providing objective information to facilitate accurate diagnosis, focused treatment and comparison for follow-up; capacity to comply with the keeping of a diary can predict likely engagement with management strategies.

Physical Examination

Special attention needs to be placed on the neurological, abdominal and perineal examination of the child presenting with symptoms of LUT dysfunction. Careful inspection of the back for bony anomaly or cutaneous manifestations of underlying spinal dysraphism should be undertaken, followed by testing of perineal and lower limb sensation and reflexes. Subtle alterations can indicate occult neuropathy [26]. Abdominal palpation can identify a distended bladder or faecal impaction and genital examination should include careful inspection of position and morphology of urethral meatus, as well as signs of dripping from an ectopic ureter in girls.

Investigations

Urinalysis may indicate clinical conditions missed on physical examination. It can alert the clinician to urinary tract infection, diabetes mellitus or renal pathology resulting in glycosuria or proteinuria. Asymptomatic bacteriuria can exist in the incontinent child as a result of dysfunctional voiding and incomplete bladder emptying. Additionally, the inflammatory sequelae of infection can give rise to overactive bladder symptoms and dysuria can promote sphincter activity during voiding, further exacerbating the underlying problem.

Uroflowmetry involves measurement of urine flow and ultrasound determination of residual urine (PVR). These tests are non-invasive and can be performed in the outpatient setting with a well-hydrated and cooperative child. Measuring urine flow during voiding gives information about bladder function and outlet function, useful in both diagnosis of disorder and evaluation of treatment impact. However, uroflowmetry is influenced by both patient cooperation and voided volume. Repeat testing in the same setting increases accuracy exponentially, with three tests recommended as the optimal balance between accuracy and time-consumption [1].

Uroflow studies need to describe four elements: volume voided, post-void residual, flow rate, flow pattern [19]. In children, the flow pattern (curve) is of greater diagnostic importance than the flow rate. The curve is determined by detrusor contractility, abdominal straining and nature of the bladder outlet. Normally smooth and bell-shaped, anomalies of flow curves can be characteristic of particular dysfunctions (Fig. 9.2). Specific terms have been developed to describe these [6], including:

- "Tower" curve is of high amplitude and short duration, indicating overactive bladder
- "Staccato" curve has sharp peaks/troughs that may indicate intermittent sphincter activity during voiding
- "Interrupted" curve shows discrete peaks with intervening zero flow segments, often indicative of voiding by abdominal straining
- "Plateau" curve is of low amplitude and prolonged duration, indicating fixed outlet obstruction

Urinary tract ultrasound is vital for all children with evidence of LUT dysfunction [19], to screen for structural anomalies and provide information about potential coexistent conditions. This scan should include detailed assessment of upper tracts (kidneys and ureters), bladder views when full and after voiding, and rectal diameter. Structural anomalies of the urinary tract will require individualised management and referral to a paediatric urologist should be considered. Full-bladder views allow assessment of bladder wall contour and thickness, abnormalities of which point to long-standing problems with bladder storage and emptying [27]. The volume of the bladder when the child feels 'full' provides some information about bladder storage ability and sensation. Post-void bladder volume should be <20 mL or <10% of expected bladder capacity. Finally, transverse rectal diameter can provide vital information on coexisting bowel dysfunction. A dimension of >3 cm without urge to defaecate is a strong indicator of faecal retention [28, 29].

Micturating cystourethrogram (MCUG, VCUG) has no place in the routine assessment of a child with urinary incontinence. When indicated for the further evaluation of identified structural anomalies presenting with incontinence, combination with urodynamic assessment in a *fluoroscopic urodynamic study* (FUDS) should be considered.

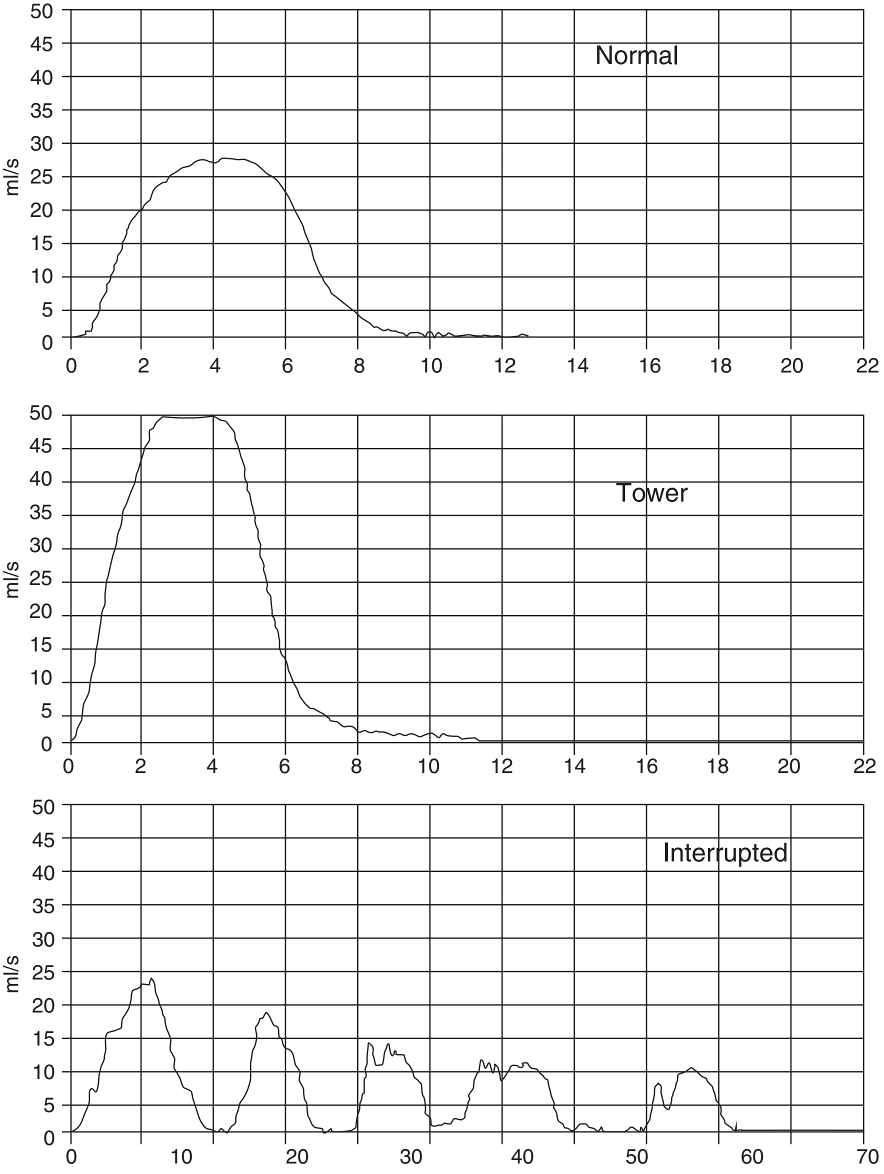


Fig. 9.2 Characteristic uroflow curves

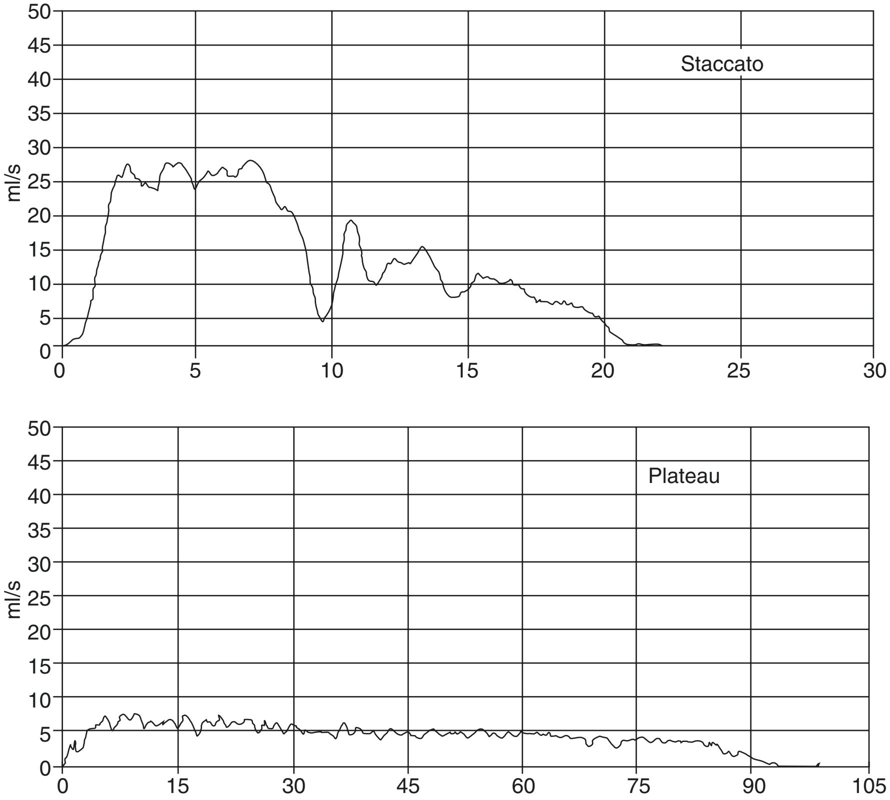


Fig. 9.2 (continued)

Clinical Conditions

There is considerable heterogeneity of symptoms in LUT dysfunction and each child should be carefully assessed as above, with decisions dependent on their individual clinical picture. Several recognisable clusters of symptoms are given specific names described here [6]. More than one can coexist in a given patient.

Overactive Bladder (OAB)

OAB is characterized by urgency, frequency and nocturia or enuresis; often accompanied by wetting. Children use typical postures such as leg crossing, ‘freezing’ and applying perineal or penile pressure (squatting on heel or ‘holding’) to inhibit urgency and postpone voiding. Investigations usually reveal frequency >8 times per day, low voided volumes, tower uroflow curve and complete bladder emptying. Invasive urodynamic studies usually show detrusor overactivity.

Voiding Postponement (VP)

Some children habitually ‘hang on’, with voiding frequency <3 times daily and often large voided volumes. They may experience incontinence from an overfull bladder and may concomitantly restrict oral fluid intake to reduce the incontinence.

Underactive Bladder (UAB)

UAB describes children who need to raise their intra-abdominal pressure by straining to initiate, maintain or complete voiding. It may be a consequence of longstanding voiding postponement or dysfunctional voiding [30]. Investigations often show interrupted flow pattern and incomplete emptying. Voiding frequency is often reduced, but may be increased due to prompt refilling of an inadequately emptied bladder. Invasive urodynamic studies usually show poor detrusor contractility or underactivity.

Dysfunctional Voiding (DV)

The child with DV habitually contracts their urethral sphincter and/or pelvic floor muscles during voiding, resulting in a staccato flow pattern and impaired bladder emptying. They typically have associated problems with bowel emptying. Vesicoureteric reflux is seen in one third to one half of these patients, likely secondary to the high voiding pressures of this condition.

Bladder Outlet Obstruction (BOO)

BOO refers to impediment of urine flow during voiding. It may be mechanical or functional, static of phasic and will be demonstrated on invasive urodynamics as increased detrusor pressure with low urine flow rate. In boys presenting with incontinence and plateau flow pattern, mechanical outlet obstruction such as posterior urethral valves or urethral stricture should be considered.

Bladder-Bowel Dysfunction (BBD)

Coexistence of bowel and bladder dysfunction is widely recognised and the importance of bowel assessment and management in the child presenting with urinary incontinence cannot be over-emphasised [31]. Some children without identifiable neurologic abnormality have coexistent LUT and bowel dysfunction that parallels that seen in children with neurologic conditions. Previous terms applied to this clinical picture include "Hinman Syndrome" and "Dysfunctional Elimination Syndrome". When severe, these children may require management approaches akin to those employed for children with known neurologic disorders.

Other

Stress incontinence involves leakage of urine due to rise in intra-abdominal pressure without detrusor contraction. It is very uncommon in children. Toilet-trained girls who experience small or moderate wetting shortly after voiding may be pooling urine due to urethro-vaginal reflux.

Extraordinary daytime-only urinary frequency (previously termed idiopathic pollakiruia) applies to a child usually 3–8 years old with extremely frequent need to void small volumes at least once per hour solely by day. Incontinence is rare and nocturia is absent. The aetiology is unknown, but may involve a sensory disturbance and/or psychological stressors [32, 33]. Investigation for diabetes mellitus or UTI should be performed. The symptoms usually resolve spontaneously after some weeks to months.

Management

Urotherapy

The principal therapeutic strategy for incontinence in children is non-pharmacologic and non-surgical. Urotherapy involves behavioural modifications tailored to the specific condition and is first-line treatment for all types of daytime urinary incontinence. As a general rule, bowel dysfunction is treated before bladder and day wetting before enuresis. Active involvement of the child is essential. '*Standard urotherapy*' includes educating the child and family about normal LUT function, providing advice on diet and fluid intake, active management of bowel dysfunction and implementing behavioural modifications such as timed voiding, avoidance of holding-on manoeuvers and optimised voiding posture. Regular supportive follow-up with encouragement and reinforcement is crucial. Treatment success is dependent on patient adherence with some centres reporting 40% cure rate with this alone [34].

'*Specific*' *urotherapy* includes pelvic floor muscle re-education +/− biofeedback, neuromodulation (such as TENS) and intermittent catheterisation. Biofeedback and neuromodulation produce generally positive results but do require particular expertise and infrastructure and are thus not widely available. Cognitive and psychological therapy may be useful adjuncts to mitigate the impact of incontinence on the child's psychosocial function, but do not often result in cessation of wetting.

Pharmacotherapy

For children refractory to urotherapy alone, pharmacotherapy is introduced. The addition of pharmacological agents should be considered if there is inadequate response to 3–6 months of optimised urotherapy with good patient engagement. Urotherapy should continue throughout pharmacologic therapy.

Acetylcholine Receptor Agents

Anticholinergic medication has remained a mainstay of treatment for urotherapy-resistant overactive bladder in children. The muscarinic ACh receptor M3 subtype regulates detrusor contraction, so blocking these receptors inhibits spontaneous detrusor contractions [35]. Adverse effects are dose-dependent and common: resulting from blockade of these same receptors in other organs. Dry mouth, flushing, concentration issues, tachycardia, constipation and blurred vision are the most commonly reported and may lead to patient discontinuing therapy [36]. Particular care should be taken to evaluate for and treat concomitant constipation as the commencement of antimuscarinic may aggravate bowel dysfunction [31]. Residual volumes should be monitored during treatment and attempted withdrawal of agent every 6–12 months should be considered [30].

Oxybutinin is the most widely used agent, available in both oral and transdermal forms. The transdermal form offers potential to avoid first-pass metabolism and reduce side effects in some, as well as providing more consistent dose delivery. However, some patients experience skin irritation from the patch adhesive, it is harder to titrate dose and not universally available. Newer agents introduced for OAB in adults are increasingly being used off-label in children. Bladder-selective agents, such as tolterodine, offer equivalent efficacy with fewer side effects [37] and M3 receptor-selective agents, such as solifenacin, promise both improved efficacy and reduced adverse effects [38].

Adrenergic Receptor Agents

Alpha-adrenergic blockers theoretically facilitate bladder emptying by blocking α-adrenergic receptors, concentrated at the bladder neck and urethra. By reducing bladder outlet resistance, voiding pressures drop and the drive for obstruction-related detrusor overactivity is removed. These agents may be useful in children with treatment-resistant dysfunctional voiding [39, 40], but treatment of choice for dysfunctional voiding remains specific urotherapy [41].

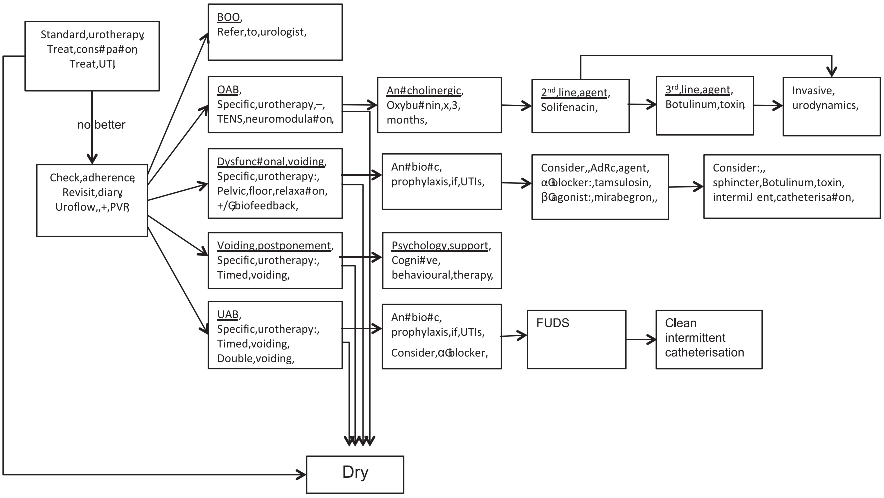
Beta-adrenergic agonists are a new class of agent for the management of incontinence. Activation of the β_3 receptor in the detrusor leads to muscle relaxation and increase in capacity. Mirabegron is the first agent in this class developed for urologic use and early evidence for role in paediatric incontinence management is emerging, either as a single agent [42] and in combination with antimuscarinic agent [43].

Botulinum Toxin (Botox)
Botulinum toxin is a potent neurotoxin that irreversibly inhibits acetylcholine release at the presynaptic junction, as well as reduces bladder sensory purinergic receptors [41]. Injected directly into the detrusor muscle, it improves bladder storage ability and compliance and reduces detrusor overactivity [44]. Most studies suggest duration of effect of 6–9 months, although in some cases it is longer. Repeat treatment seems equally efficacious, although longterm impact of repeated injections on the bladder wall is not yet known. Risk of urinary retention and need for intermittent catheterisation following treatment is low but not trivial [41]. Despite the invasive nature of administration, botulinum toxin provides a valuable treatment of last resort for a child with persistent incontinence due to refractory bladder overactivity.

Surgical

Surgery may be necessary where structural genitourinary anomalies are identified as the cause of the incontinence. Boys with bladder outlet obstruction due to urethral stricture or posterior urethral valves will require surgical intervention. Girls with continuous incontinence resulting from ectopic ureter may likewise require surgical cure.

Therapeutic Algorithm



Conclusion

Lower urinary tract dysfunction in children is common. Most patients have no structural, neuropathic or other organic cause found. Considerable comorbidity exists with concomitant bowel dysfunction, nocturnal enuresis and recurrent UTIs. Inadequately managed, it gives rise to significant psychosocial morbidity. The initial diagnostic approach consists of a focused medical history and physical examination, bladder diary and urinalysis. Clinic-based investigations include urinary tract ultrasound, uroflowmetry and measurement of post-void residual volumes. Urotherapy is a first-line treatment for most conditions related to incontinence, but pharmacological treatment may also be helpful. Initial treatment for most children can be provided by a primary care physician. Refractory cases and those in whom a structural cause is suspected benefit from specialised treatment in a multidisciplinary service involving paediatricians, continence physiotherapists, psychologists and urologists. Paediatric clinical trials comparing the efficacy of various urotherapeutic interventions are lacking, but significant advances in treatment options have been made in recent years. Key contemporary publications in this field [31, 34, 41, 45] could be read for further detail.

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Chapter 10
Disorders of Continence: Enuresis

Nathalie Webb, Janet Chase, Marjorie Burgess, and Felix Grusche

Key Points
- Enuresis, also termed bedwetting or nocturnal enuresis, is a common disorder in children, with significant potential for adverse psychosocial impact
- Early management of enuresis is important for social development and self-esteem of the child
- The primary goal in treating enuresis is to teach the child to self-awaken to the sensation of a full bladder and independently void in the toilet
- Enuresis alarms are associated with the highest cure rates and lowest relapse rates of any single treatment option
- Combination therapy (enuresis alarms and pharmacologic management) may increase cure rate

Introduction

Definitions

Enuresis (nocturnal enuresis, bedwetting) is involuntary, intermittent wetting during sleep in a child ≥5 years old who is neurologically and developmentally normal. It is subclassified into:

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- *monosymptomatic* nocturnal enuresis (MNE) describing nighttime wetting in children without any other lower urinary tract symptoms
- *non-monosymptomatic* nocturnal enuresis (non-MNE) describing night wetting in children with complicating organic factors or other lower urinary tract symptoms, such as dysfunctional voiding or daytime urinary incontinence

Primary enuresis is used for children who have never achieved nighttime dryness for >6 months; *secondary* enuresis applying to those who have previously been dry at night for ≥6 months [1].

Prevalence

Enuresis is a common condition occurring in 5–10% of 7-year olds, with boys affected more than girls [2]. Without intervention, prevalence decreases with age by about 15% per year, but enuretic problems may persist in 1–2% of adults; often with greater severity and frequency [3, 4] suggesting spontaneous resolution occurs mostly at the milder end of the symptomatic spectrum [5].

Pathogenesis (Cause)

Enuresis is a heterogeneous condition, with several likely underlying pathophysiological mechanisms leading to a mismatch between nocturnal urine production and bladder storage ability and simultaneous failure of conscious arousal in response to the sensation of needing to void [5] (Fig. 10.1).

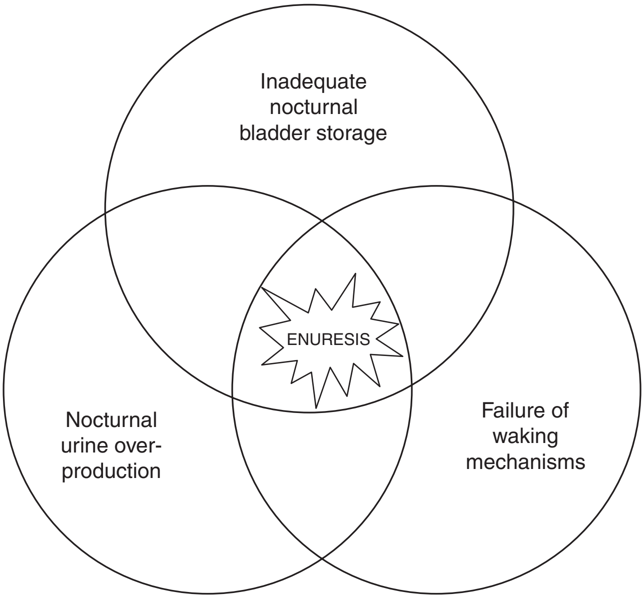

Inadequate nocturnal bladder storage
ENURESIS
Nocturnal urine over-production
Failure of waking mechanisms

Fig. 10.1 Interplay of factors contributing to enuresis

Despite extensive research, the exact pathophysiology remains ill-defined. The traditional concept of maturational delay accords with the rate of spontaneous resolution, although multifactorial influences on bladder storage, urine volume and arousal responses are all implicated.

It is well recognized that enuresis may have a profound psychological and social impact on the affected child and their family [6]. The reversal of these impacts with successful cure of enuresis provides strong indirect evidence that the psychological and behavioural problems are the effects rather than the cause of enuresis [7].

Genetics

Research has demonstrated a higher incidence of enuresis in children with a family history of bedwetting, with 60–80% of all enuretic children having affected relatives [8]. The inheritance pattern appears to be autosomal dominant and geneticists have identified possible candidate markers on chromosomes 12, 13, and 22. In addition to the multiplicity of possible genes, genotype and phenotype correlate poorly, so individuals within a family may well have considerably different underlying mechanisms, response to treatment and prognosis.

Sleep and Arousal

Contrary to traditional thinking, enuresis is not caused by deep sleep. Children with enuresis have normal polysomnographic patterns, but are characteristically very difficult to wake from sleep [9]. Cystometric studies performed on children during sleep have shown that they have more difficulty arousing from sleep during non-rapid eye movement phases, which predominately occur during the first two-thirds of the night [10]. Elevated arousal threshold renders the child unable to wake up in response to the urge to void, and the micturition reflex follows. This observation has led to the theory of slow-to-mature sleep arousal mechanism at brainstem level as a cause of enuresis [9].

Alternative theories, especially pertinent to those with non-monosymptomatic enuresis (non-MNE), suggest that presence of frequent uninhibited bladder contractions during sleep may generate similar sleep disturbance to that seen in sleep apnoea. Brain prioritizing of sleep integrity elevates the arousal threshold to mitigate constant stimuli from airways or bladder. Subsequent daytime impact of disturbed sleep; such as sleepiness, decreased performance and headache are common in enuretic children [10].

Reduced Bladder Storage Function

Children presenting with *monosymptomatic* enuresis (MNE) are often assumed to have normal underlying bladder function, however many recent studies attest to a pathogenic role of occult bladder overactivity [11, 12]. Uninhibited detrusor contractions cause the bladder to signal need to void at much less than age-expected bladder capacity (see Chap. 11, Fig. 10.1). Storage inadequacy of a child's bladder can be reflected in daytime voided volumes [13]. A bladder diary recording several days' voids, but omitting the first morning void, can be used to determine an individual's average volume for comparison with expected-for-age value. While the child may not report daytime problems, the diary often also demonstrates diurnal urinary frequency. Children with functionally reduced bladder storage usually have *primary* enuresis, wetting the bed every night and/or having several wetting episodes per night.

Polyuria and ADH Secretion

Production of nocturnal urine volume in excess of bladder storage capability will result in either nocturia or enuresis [5]. Nocturnal polyuria may be implicated in both primary and secondary enuresis, even when bladder storage is not reduced. Normally, antidiuretic hormone (ADH) levels increase at night causing the kidneys to concentrate urine, reducing volume to 'fit' so that the child neither gets up to void nor wets the bed. Studies [14, 15] have demonstrated derangement of ADH circadian rhythm resulting in nocturnal polyuria in many enuretic children. This derangement may vary from night to night.

Polyuria is also seen in several disease states including diabetes mellitus, diabetes insipidus and sickle cell anemia. It can also occur in children who drink excessive amounts of fluid in the evening or wake to drink overnight.

Psychological/Behavioral

Psychological stressors alone do not cause enuresis, but may play a role in development and complicate treatment. 20–30% of enuretic children have clinically-relevant concomitant psycho-behavioural disorders [16]; potentially linked in several ways:

- the psychological disorder may be a consequence of the incontinence
- the psychological disorder may potentiate a genetic predisposition to enuresis
- incontinence and the psychological disorder may both be due to a common neurobiological dysfunction eg ADHD
- the two conditions may coexist by chance

Unquestionably, enuresis can have profound psychosocial impact on the child and family. In general, these children have lower self-esteem and more anxiety, depression, attention problems and aggressive tendencies than age-matched controls [2]. Children with non-MNE have higher rates of psychosocial morbidity than those with MNE, and secondary enuresis is associated with a higher incidence of stressful life events and psychological problems. Treatment of the enuresis can generate a marked reversal of the adverse psychosocial and behavioural issues [5].

Food Sensitivity

Diet may also play a role in enuresis, although scientific evidence is lacking. Dairy products contain tryptophan, potentially increasing serotonin levels and leading to decreased sleep arousal. Carbonated and caffeinated beverages increase urine production and acidic citrus fruits and spicy foods can cause bladder irritability, causing more bladder contractions. Exclusion diets have not shown reliable impact on enuresis resolution, however.

Non-monosymptomatic Enuresis: Overactive Bladder, Dysfunctional Voiding, Constipation

Children without daytime wetting may still have significant bladder overactivity manifesting as frequency and urgency, which is often unrecognized. Overnight, the lack of conscious management may result in failure to suppress urge sufficiently to reach the toilet.

Children with lower urinary tract dysfunction (see Chap. 11) that results in incomplete emptying may have reduced capacity to store additional urine. Furthermore, incomplete bladder emptying risks urinary tract infection (UTI), with resultant bladder irritation and impaired bladder function.

Irregular and infrequent stooling can also affect the ability of the bladder to fill and empty properly. A full rectum can put pressure on the bladder, preventing it from storing or emptying well. During bladder filling, expansion of the bladder may be restricted by a full rectum. Additionally, when the rectum chronically intrudes upon the bladder, it can decrease the sensation of bladder fullness. These elements are particularly important to consider in a child with secondary enuresis (Table 10.1).

Table 10.1 Causative conditions for non-MNE and/or secondary enuresis

Condition	Mechanism
Constipation	Reduced bladder storage
Urinary tract infection (UTI)	Reduced bladder storage Overactive bladder (OAB)
Diabetes (mellitus/insipidus)	Nocturnal polyuria
Sleep-disordered breathing	Impaired arousal
Neurogenic bladder	Reduced bladder storage Neuropathic detrusor overactivity

Investigations

History

A thorough history is essential in the evaluation of enuresis. While most school-aged children can report school-day voiding pattern, this may not be reliable [5]. A bladder diary, recording time and volume of voids as well as associated fluid intake, incontinence episodes, urgency and bowel movements provides objective information and attests to the child and family's motivation to engage in management. It also helps identify those children with occult daytime bladder dysfunction who may purposefully reduce daytime fluid intake to prevent daytime incontinence episodes. These children may take in large volumes of fluid in the evening, driving high night-time urine production. Since bowel and bladder function are closely interrelated, concomitant constipation must also be sought and treated [17].

Physical Examination

The physical examination should focus on the bladder and bowel, including assessment of genital and neurological systems. The abdomen should be palpated for stool and bladder distension. The spine should be examined for cutaneous defects, hair tufts or pigmentation that may indicate occult spinal anomalies. Genital exam should include urethral position, meatal stenosis and signs of abuse or trauma. Uroflowmetry or observation of urinary stream should be considered if it sounds abnormal on history. Assessment of lower limb strength, tone, reflexes and sensation should be performed seeking underlying neuropathy.

Laboratory Tests

A urinalysis should be conducted in every child with enuresis to evaluate for glycosuria (diabetes mellitus), low specific gravity (diabetes insipidus) and potential infection. If symptoms of UTI are present or urinalysis is positive for leucocytes or nitrites, a urine culture should also be performed.

Imaging Studies

Radiographic studies are not routine for children with enuresis alone, but may be helpful in children who also have daytime symptoms. Ultrasound of the urinary tract identifies morphologic anomalies and can also evaluate bladder wall thickness and post-void residual volumes. Other investigations are only indicated in cases of suspected urinary tract obstruction or neurogenic bladder.

Treatment Modalities

Just as there is no simple cause for bedwetting, there is no single solution. Children with associated daytime incontinence, urgency and/or constipation will need to have these symptoms addressed before night time therapy can begin.

Conventional enuresis management approaches include behavioral modifications, alarm therapy and pharmacologic treatment. These interventions should be chosen based on the type and severity of symptoms, the family's goals for treatment and the benefits and disadvantages of the management modality. A vital consideration is the child's self-esteem. Enuresis can improve over time with maturational development, however treatment should not be deferred on that basis alone [17], as spontaneous resolution in children over the age of 5 years is low and slow (15% per year [2]). All enuretic children, regardless of age and severity, deserve evaluation and intervention guided by the degree of concern and motivation of the child.

Behavioral Therapy

'Basic bladder advice', sometimes termed standard urotherapy (see Chap. 11), involves information and demystification, management of any bowel dysfunction, regulation of daytime voiding frequency and sound drinking habits with liberal fluid intake early in the day and restriction in the evening [17]. A normal varied diet is advised, although for some children, a high protein and sodium load may lead to a later osmotic diuresis. It is vital that the child and family understand the causes of enuresis; most especially that it is not intentional and should not be punished. While recommended and widely practised as first-line treatment, there is no evidence that basic bladder advice will actually hasten the resolution of monosymptomatic enuresis [1]. However, the discussion involved in the process provides an opportunity for child, family and clinician to develop to a shared understanding of options and goals for treatment.

Simple behavioural interventions include fluid restriction, lifting or timed wakening and reward systems. These may achieve some reduction of wetting episodes, but evidence for curative impact is small [18].

Alarm Therapy

Alarm therapy is designed to help the child learn to wake to sensation of a full bladder. Used correctly and consistently, alarm intervention results in dryness for about two-thirds of treated children [19]. It is presumed to cure enuresis by conditioning effect on arousal, but has also been shown to increase nocturnal bladder storage [20, 21]. Alarms have been extensively studied, with Grade 1a evidence base to support their use [1], however they do require significant motivation on behalf of parent and child to optimise impact [22]. While there is suggestion that body-worn alarms are more acceptable than traditional pad-and-bell systems, there is insufficient evidence of superiority of cure with one type over others [23].

Engagement of both child and parents is vital to the success of alarm therapy. Parents may need to be reminded that learning is progressive and positive reinforcement is very important, especially during the first few weeks of treatment [2]. They may also need to consider modifying sleeping arrangements to best support the learning process, as it may be necessary for parents to wake the child, help them use the toilet and change the bed. The alarm should not be turned off until the child is awake and standing on the floor. The child will gradually learn to awaken to the alarm without reminders or parental help. Finally, the child will use this new sensation to wake himself and use the bathroom without the use of the alarm. This process takes 2–3 months on average but may take up to 6 months. Initial failure does not preclude future successful treatment so this option may be revisited as a treatment option when the child is older.

Pharmacologic Therapy

Medications used in the treatment of enuresis include desmopressin, anticholinergic and tricyclic agents.

Desmopressin

Desmopressin acetate (or DDAVP) is a synthetic analogue of anti-diuretic hormone (ADH, vasopressin). The anti-diuretic activity results in concentration and thus reduced volume of urine produced after dosing, even in those with apparently normal endogenous ADH levels. For enuretic children, this allows them to sleep dry or wet less at night. It also has grade 1a evidence of effect [24], with estimated 30% of children responding completely and a further 40% partial responders. Curative potential is low with >90% relapse rate after short-term treatment [25]. It can be particularly beneficial for social events, such as sleepovers or overnight camps.

The main risk is of water intoxication and hyponatraemia if excessive fluid intake occurs after dosing. This is avoided by stopping fluids 2 h prior and for 8 h after dosing. Desmopressin is available in oral tablet, nasal spray and sublingual wafer form. Previously used widely, the nasal spray is no longer recommended for enuresis treatment due to inconsistent absorption rates and longer half-life leading to apparently greater risk of hyponatraemia [26]. The sublingual ‘melt’ formulations are 120 μg or 240 μg and should be taken 30–60 min before bed. Bladder emptying immediately before bed is mandatory.

Desmopressin can be prescribed in combination with an enuresis alarm. Most children who wet the bed do so within the first 90 min to 2 h after going to sleep, coinciding with non-REM sleep. By adding desmopressin to the enuresis alarm therapy, the wetting episode may be shifted to early morning when the child may be more responsive to alarm waking. After a period of time, the medication can be reduced and then discontinued.

Anticholinergic Therapy

Anticholinergic agents, such as oxybutynin, are effective in the treatment of enuresis [27]; especially when there is evidence of reduced bladder storage, frequency and/or urgency. Oxybutynin works by relaxing the smooth muscle of the bladder, allowing more urine to be stored and delaying the urge to void. Side effects include constipation and increased post-void residual volumes. These need to be carefully watched for, as they can exacerbate enuresis. For children without daytime symptoms in whom nocturnal bladder overactivity is suspected, single dose administration at bedtime may be sufficient.

Tricyclic Agents

Imipramine and other tricyclic antidepressants have been used since 1960s to treat enuresis, with evidence of positive impact in the short term [28]. The mechanism of action is anticholinergic effect increasing bladder storage combined with a noradrenergic effect decreasing the excitability of the bladder. Lack of sustained impact after treatment cessation and the risk of potentially serious adverse effects, including death with overdose, has resulted in it no longer being recommended for treatment of enuresis, unless all other therapies have failed [17].

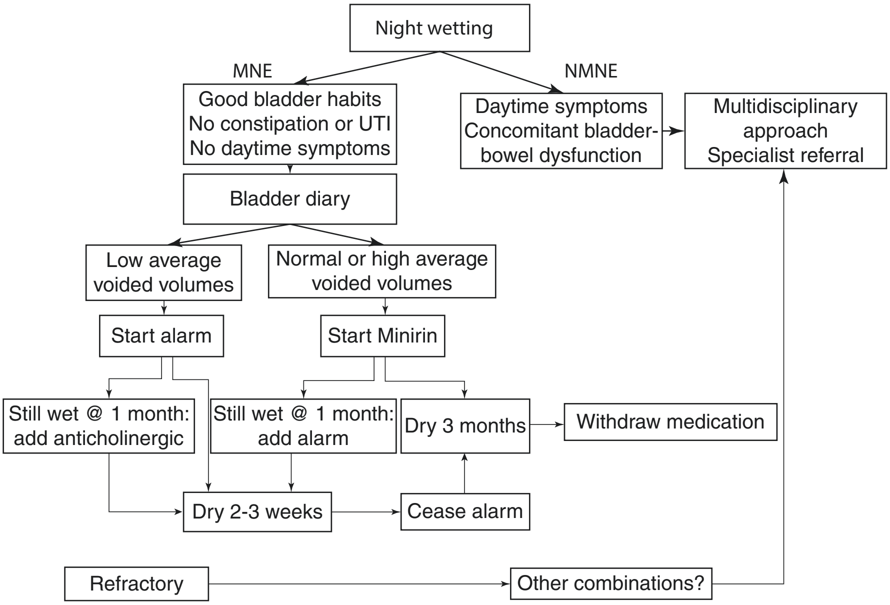
Botulinum Toxin

The use of botulinum toxin in the treatment of refractory bladder overactivity has a growing evidence base. Injected directly into the muscle, the primary effect of botulinum toxin A appears to be inhibition of release of acetylcholine, ATP and substance P [29]. It results in a profound and long lasting inhibition of the mechanisms thought responsible for overactive bladder symptoms [30]. The resultant increase in bladder storage and inhibition of detrusor contractions improves diurnal frequency and urgency as well as enuresis.

Alternative Treatment

Complementary treatments such as hypnosis, psychotherapy, acupuncture, chiropractic and medicinal herbs have all been used to treat enuresis, however review of trials provides no evidence to support these, apart from weak evidence for acupuncture. There is no reliable information comparing complementary methods with methods proven to be effective such as alarms and desmopressin [31].

Treatment Algorithm



Conclusion

Enuresis or bedwetting is a common childhood condition that can cause inconvenience, stigma and stress. Once the child demonstrates a desire for dryness, initiating management is imperative to reduce adverse impact on the child's self-esteem and social development. A heterogeneous spectrum of disorders, imbalance among sleep arousal, functional bladder volumes and nocturnal urine production is implicated. Focused history and examination can help define appropriate therapeutic approach. Overall, alarms offer the best chance of cure. Desmopressin can be added for patients with apparent nocturnal polyuria. Adjunctive anticholinergic therapy should be considered for those with functionally small bladders. Regardless of treatment approach, the provider, child and the family need to negotiate a management plan including a system for measuring progress and regular follow-up visits.

Guidelines

The International Children's Continence Society (ICCS) has published recommendations for the evaluation and treatment of enuresis [17, 30, 32].

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Chapter 11
Constipation

J. Christopher Austin

Introduction

Key Points
- The majority of children who present with urinary tract infections and problems with daytime and nighttime incontinence will have associated bowel symptoms such as constipation and encopresis (fecal incontinence).
- In children presenting with the complaint of constipation up to 2/3 will also have associated symptoms of bladder dysfunction
- About 95% of patients with constipation have functional constipation and will respond to therapy with behavior modification (timed evacuation after meals) and laxative therapy.
- The treatment of constipation will improve or cure some children with daytime incontinence and recurrent urinary tract infections.

Constipation is commonly seen in pediatric patients. It is a frequent complaint seen by pediatric gastroenterologists as well as primary care providers. Constipation is very relevant to pediatric urology patients as it is seen frequently in patients presenting with urinary tract infections and/or voiding dysfunction. *Dysfunctional elimination* is the combination of bowel and bladder dysfunction that was initially described in patients with recurrent urinary tract infections and vesicoureteral reflux. *It is important to seek out symptoms of bowel dysfunction in patients seen with urinary complaints and vice versa*. The treatment of constipation is an integral part of the treatment regimen in children with voiding dysfunction. Encopresis, or fecal incontinence associated with constipation and chronic fecal impaction may also be

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present in patients with voiding dysfunction and recurrent urinary tract infections. In children with constipation an organic cause should be sought out during the evaluation including neurologic, endocrine, or anatomic causes. In 95% of patients no etiology is found and they are classified as having functional constipation.

Definition of Constipation

There are multiple definitions of constipation. The Rome III definition of functional constipation is listed below. This is intended for children older than 6 months of age. The strict Rome criteria require symptoms to be present for longer (1–2 months) period of time but recent guidelines suggest that 2 weeks is a sufficient duration of symptoms to make the diagnosis.

Rome III Definition of Functional Constipation

Two or more of the following for at least 2 weeks

- Two or less defecations per week
- At least 1 episode of fecal incontinence after toilet training
- History of excessive stool retention
- History of hard or painful bowel movements
- History of large-diameter stools that may obstruct the toilet
- Presence of a large fecal mass in the rectum (abdominal exam, rectal exam, or radiograph).

Functional constipation is present only if there is an absence of some underlying medical condition that predisposes the child to constipation. This is commonly seen in children with developmental delay, neurologic disorders or prematurity. *Given the relatively benign nature of the treatment of constipation with stool softeners or laxatives, treating patients with bowel or urinary complaints should be considered when the diagnosis is suspected.*

Evaluation

The first step in evaluating a child with constipation is a thorough *elimination history*. This should include whether the child passed meconium within 48 h of birth (delay in this is worrisome for Hirschprung's Disease), the frequency, size and consistency of bowel movements. The presence of associated symptoms such as abdominal pain, vomiting, blood in the stool, withholding bowel movements, fever, and abdominal distention should be noted. The grading of bowel movements using an objective scale such as the Bristol stool scale or Lane Scale (modified Bristol scale for children) can be useful confirming if the child is passing hard stools. Large diameter stools or stools which clog the toilet are signs of constipation as well. The history should probe for possible organic causes of constipation such as symptoms starting at under 1 month of age, hypothyroidism, botulism, neurological disorders, medication side effects, and failure to thrive. If there are alarming signs and symptoms present the child doesn't have functional constipation and should be further evaluated by a specialist. There is a high incidence of concomitant bladder and bowel symptoms and thus the frequency of urination, presence of urgency, posturing, daytime or nighttime incontinence, and urinary tract infections should be sought.

A *physical exam* should be performed with emphasis on the abdominal (palpable stool, tenderness, abdominal distention, anal position and appearance), neurological (reflexes, strength, anal wink and perineal sensation), and back exam (sacral dimple, lipoma, or hair tuft). Any suspicious neurological findings should lead to evaluation for a spinal cord abnormality such a tethered spinal cord. A rectal exam is not considered mandatory to diagnose a child with functional constipation but may be useful in detecting a fecal impaction if suspected and evaluating for an anorectal anomaly. Abdominal films have been used to assess the fecal load and evaluate the spine for an occult dysraphism in the past. *It is currently not recommended that x-rays be used in the routine diagnosis of constipation*. Abdominal radiographs may be useful of the diagnosis of fecal impaction when the abdominal exam is not felt to be reliable, such as obesity or an uncooperative patient. Patients with no suspicious findings in the history and exam likely have functional constipation and should be treated.

Treatment of Constipation

The initial therapy for constipation consists of behavior modification (sitting to have a bowel movement for 5–10 min shortly after meals) and laxative therapy. A diet with normal amounts of fiber and fluid intake should be followed. Studies of additional fiber supplementation have not shown efficacy in the treatment of constipation and thus increased fiber is not recommended as first line therapy. If there is suspicion for fecal impaction (palpable stool on exam, large hard stool ball on rectal exam, or findings of excessive stool in the lower colon on a plain radiograph) a bowel cleanout to disimpact the rectum should be done before treating. This was historically done with enemas but currently oral laxative (PEG 3350) is recommended as the first line treatment. Once disimpaction is complete if needed then the child is maintained on laxatives to keep the stools soft and allow the chronically over distended rectum to regain its normal tone and function. There is good evidence for the efficacy of laxative and stool softener therapy in constipation. For maintenance therapy PEG 3350 has been shown to be superior to other oral treatments. The chronic uses of enemas is not recommended in patients with functional constipation. The most common treatments and their usual dosages are listed below.

The maintenance therapy should be continued for a duration of at least 2 months before weaning and only if the symptoms of constipation present have been fully treated. If the child has difficulty with fecal incontinence during therapy it needs to be ensured that they are adhering to the scheduled bowel evacuations and not trying to hold their stools. Of the following agents PEG 3350 is recommend as the first line therapy. Lactulose and milk of magnesia would be considered the best alternatives if the child is unable to be treated with PEG 3350.

- *PEG 3350* (Miralax™, Glycolax)- Effective dose for constipation has been reported in the 0.5–0.84 gm/kg per day given daily. Total dose may be divided in half and given twice daily which can help improve the stool consistency. The powder may be mixed with any good tasting fluids. The solution should be mixed with 17 gm of powder (filled to the mark inside the cap) into 240 ml (8 ounces) and the appropriate amount of this dilution taken (i.e.- a dose of 8.5 g would require drinking 120 ml (4 ounces) of the solution). The mechanism of action of PEG 3350 is an osmotic agent which retains water in the stool to soften it and increase the frequency of bowel movement. The dose can be adjusted up or down every 3–5 day by an ounce until the desired stool consistency is reached.
- *Milk of Magnesia* (Magnesium Hydroxide)- Effective dose for treating constipation is 1–2 ml/kg per day given in a single or divided into a twice daily dose. The maximum recommended amount is 60 ml per day. Compliance with daily doses is less than with PEG 3350. It can be mixed or flavored with applesauce, chocolate syrup, or other flavorings to improve the taste. Its laxative effect is due to a hyperosmotic effect drawing fluid into the stool and the stimulation of increased peristalsis in the small intestine.
- *Lactulose*- Supplied in a solution of 10 g/15 ml. The mechanism of action due to the synthetic dissacharide being degraded in the colon by bacteria producing lactic acid, acetic acid, and formic acid, which increase the osmotic pressure drawing water into the stool softening it.
- *Senna*- A laxative that stimulates peristalsis and is available as both oral syrup (218 mg/5 ml) and a tablet (167 mg). For children 1–5 years of age the recommended dose is 2.5–5 ml of syrup (109–218 mg) given at bedtime. For children age 5–15 years old a dose of 5–10 ml of the syrup can be used initially. Alternatively 1–2 tablets can be given at bedtime. Senna should only be used short term.
- *Mineral Oil*- Dosage for children 6–12 years old is 5–15 ml per day. After age 12 years 15–45 ml per day. Not recommended for younger children due to the risk of aspiration. The oil helps soften stools by coating them and the intestine to reduce the colonic absorption of water and lubricate hard stools to facilitate passage.

Indications for Referral

Patients with constipation that is suspected to be due to an organic cause based on the history or physical exam should be referred to a specialist for further testing which may include rectal manometry, barium enema, or spinal MRI. Patients with persistent symptoms after adequate treatment for 3–6 months should be referred to pediatric gastroenterologist.

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Chapter 12
Abdominal Pain: Urological Aspects

Carolina Acuña and Pedro-Jose Lopez

Key Points
- Abdominal pain is a common symptom in pediatric practice.
- Different pathologies may cause abdominal pain including a number of urological conditions.
- It is relevant to have these possible conditions in mind when studying pediatric patients.
- This chapter will review some of these pathologies and their initial management.

Introduction

Abdominal pain is one of the most common symptoms in pediatric practice. Aetiologies behind it are multiple; a number of urological conditions present themselves with abdominal pain that may be classified as follows:

(a) Obstructive
(b) Non obstructive pathologies: infectious diseases, malignancies and gynecological conditions

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Obstructive Urological Pathologies

1. The most classical of them all is undoubtedly urolithiasis. Nevertheless, the classical acute abdominal or flank pain that characterizes this pathology in adults occurs in only 50% of children. Preschool children are rather asymptomatic and are usually diagnosed after a UTI. Other clinical manifestations are haematuria, dysuria and/or urinary frequency. Evaluation of these patients include a complete clinical history with emphasis on dietary habits, metabolic evaluation and, ideally, stone analysis. Diagnosis in children is generally achieved with ultrasound, which may detect stones at the renal pelvis, ureteropelvic junction, proximal and distal ureter; nonetheless, it does not visualize stones at the mid ureter. Signs due to obstruction may suggest the presence of urolithiasis. In older patients plain abdominal X-ray may be diagnostic. Non enhanced abdominal computed tomography is the gold standard in the diagnosis of this pathology, although a relevant issue in children is the possible risk of accumulated radiation exposure. Therapeutic alternatives are multiple; expectant approach, extracorporeal shockwave lithotripsy and surgery. In turn, surgery offers various approaches, especially minimally invasive surgery with endourologic procedures such as ureteroscopy, percutaneous nephrolithotomy and laparoscopic procedures.

	Population	Prime symptoms	Initial approach
Kidney stones	More frequent in smaller children	Unspecific, recurrent abdominal and flank pain	Initial study of these patients should include urine analysis and an ultrasound. If US is positive, metabolic study[a] is the next step
Ureteral stones	More frequent in school age and adolescents	Acute flank pain and haematuria	
Distal ureteral stones and bladder stones	More frequent in adolescents	Acute abdominal and flank pain, haematuria and dysuria	

[a] Metabolic study: 24 h excretion of calcium, uric acid, oxalate, citrate, cysteine and sodium

Patients with calculi passed or retrieved, patients with non – glomerular haematuria and dysuria with abdominal / flank pain and patients with abdominal or flank pain without other symptoms but with positive family history or urolithiasis should be studied with an ultrasound. Patients with positive ultrasound should be referred and undergo metabolic study, specific imaging when necessary and specific treatment. Patients with ultrasound that does not identify a stone but describes urinary obstruction will need further imaging. Patients with a negative ultrasound but high suspicion of lithiasis either because of family history or urinary symptoms should also be referred. Ultrasound should be repeated in a couple of months and probably will end in metabolic study too (Figs. 12.1, 12.2, 12.3, 12.4, 12.5, and 12.6).

Fig. 12.1 Palpable abdominal mass in a 4 year old patient, who is studied for abdominal pain and constipation

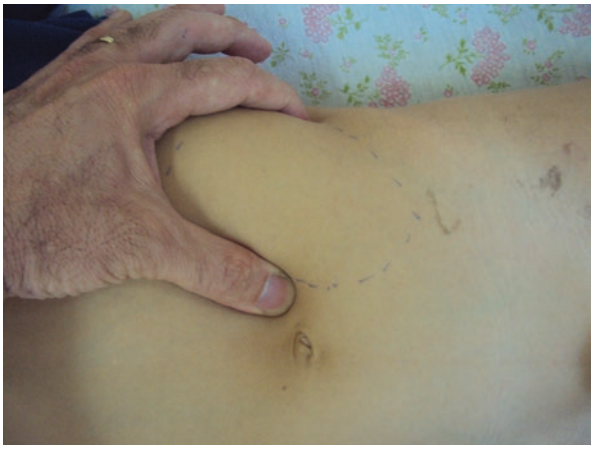


Fig. 12.2 Abdominal CT from same boy showing important mass with no renal tissue observed: diagnosis of Right PUJ obstruction

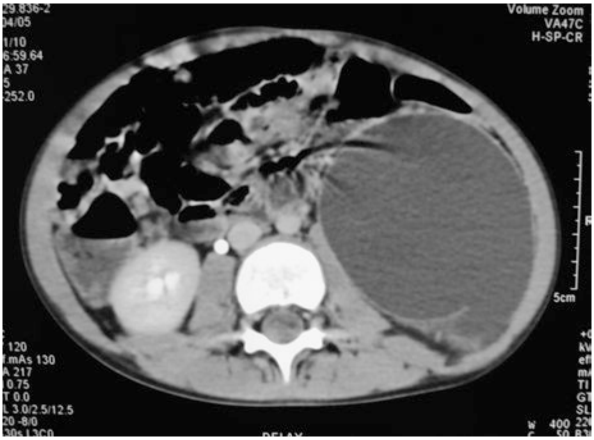
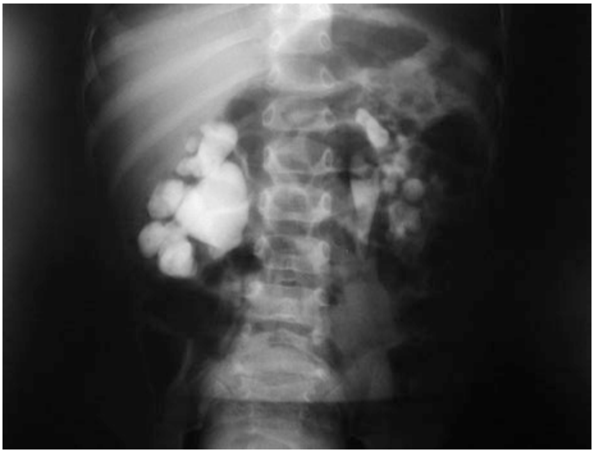


Fig. 12.3 Urolithiasis: multiple radio opaque stones in both kidneys



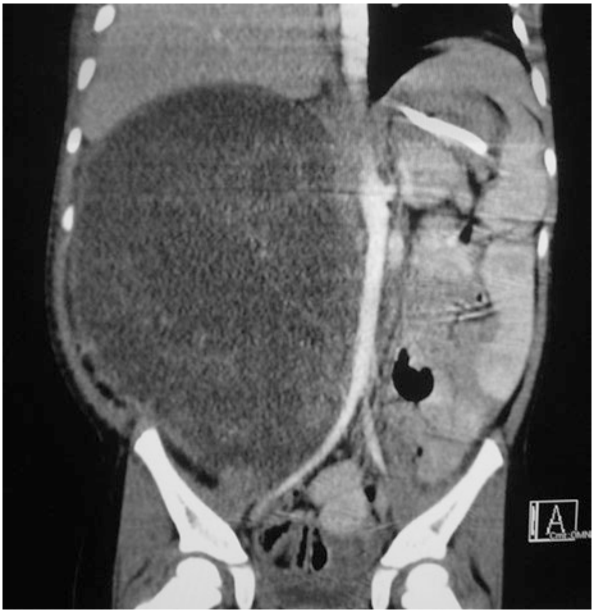


Fig. 12.4 MRI: Large heterogeneous mass at the right flank showin a kidney tumour

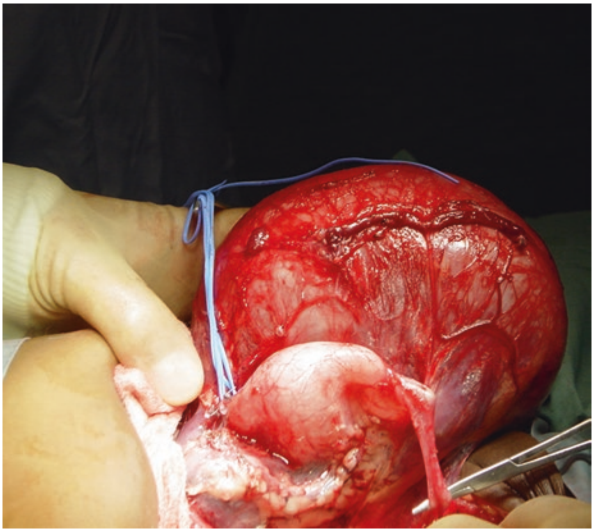


Fig. 12.5 Same Wilm's Tumour from Fig. 12.4 at surgery

2. Pelvicureteric junction (PUJ) obstruction is the most common congenital urinary obstruction. Before the spread of prenatal diagnosis the most common presenting symptom in patients with PUJ obstruction was abdominal pain and abdominal mass followed by UTI and haematuria. Nowadays, most patients with PUJ are diagnosed prenatally and are studied and "solved – operated?" before they present symptoms. Initial study should include ultrasonography, which may show the dilation caused by the PUJ obstruction, evaluate renal parenchyma

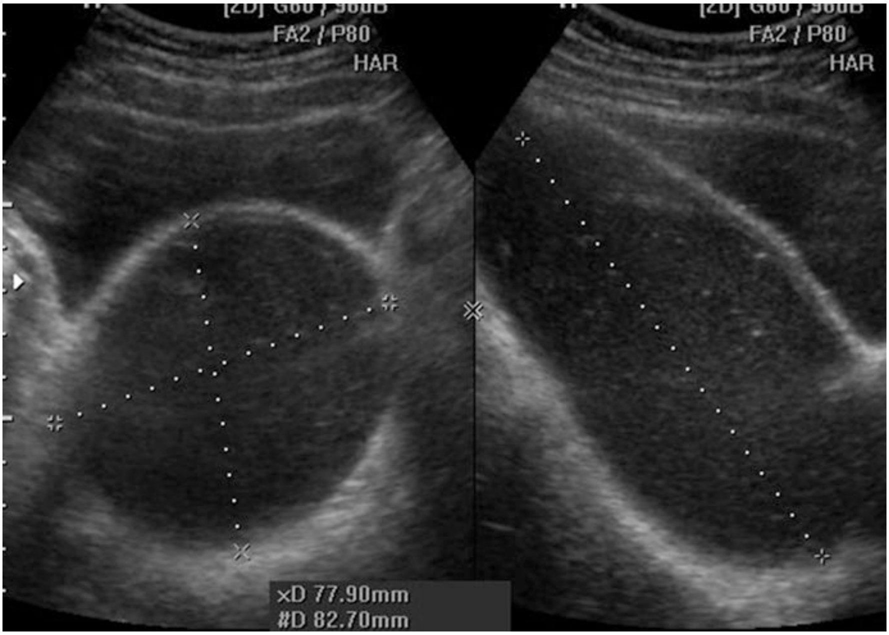


Fig. 12.6 Ultrasound: Hipoechoic fluid content distending the uterus and vagina

thickness, and suggest if renal dysplasia exists. Dynamic renal scintigraphy is very useful in the assessment of possibly obstructed kidneys. Some of these obstructions are non-progressive and asymptomatic making relevant the need to determine which kidneys will require a surgical management. Also, in these patients, you should not discard concomitant problems like vesicoureteral reflux or urinary stone disease, especially in adolescents. The surgical repair consists of a pyeloplasty either open or laparoscopically approached, which has a high rate of success and a low rate of complications.

3. The wide ureter or megaureter is caused by either obstruction or reflux occurring at the ureterovesical junction. They can be classified in primary, ureteral defects, and secondary, defects whose “cause” is extraureteral. Megaureters, especially primary ones, generally are suspected in prenatal ultrasound. Retrovesical ureteric diameter $\geq$ 7 mm from 30 weeks gestation onwards is considered abnormal and should be followed after birth. Until definitive diagnosis is made it is advisable to start antibiotic prophylaxis at least during the first 6 months. Postnatal diagnosis is usually made while studying a patient with UTI, haematuria, abdominal mass or cyclic abdominal pain. Work up must include ultrasound, voiding cystourethrogram, dynamic nuclear renography and, in specific cases, cystoscopy. Bladder outlet obstruction and vesicoureteral reflux should be studied and excluded in these patients. When anatomy is unclear, an enhanced computed tomography should be requested in order to determine the specific cause of the wide ureter and plan the appropriate surgical approach.

4. Urinary tract obstructions at other levels, such as bladder outlet and urethra, have different clinical manifestations. They may sometimes include abdominal pain but clearly not as a prime symptom.

Non-obstructive Urological Pathologies

1. Urinary tract infections are the most prevalent bacterial disease during the first 3 months of life and accounts for almost 10% of febrile episodes in infants. Clinical presentation is variable and very nonspecific, especially considering age, gender, pathogen and associated anatomical malformations. Pyelonephritis is generally characterized by fever, abdominal and or flank pain, dysuria, frequency and haematuria. Symptoms of peritoneal irritation may be present. Lower urinary tract symptoms include dysuria, frequency, urgency, haematuria and abdominal/suprapubic pain. Diagnosis is made with a properly obtained urine specimen; an ultrasound during the acute phase is very useful in confirming diagnosis and identifying an obstructive uropathy. Eventually a DMSA is useful demonstrating acute pyelonephritis and secondary scarring.
2. Renal tumours also may present with abdominal pain. The most common primary malignant renal tumour of childhood is Wilm's tumour and the most common benign solid tumour is mesoblastic nephroma. Generally renal tumour presentation is through palpable abdominal mass. Additional findings include abdominal pain, haematuria, and sometimes pyelonephritis among others. Diagnostic evaluation should include routine laboratory tests, tumour markers, imageneologic studies for accurate staging and in search for metastasis. These patients should be referred to the specialist as soon as possible.
3. Symptoms that may lead you to suspect vesico ureteric reflux (VUR) in a paediatric patient are generally related to urinary tract infections. In newborns and pre-school patients symptoms are vague such as irritability, failure to thrive, poor feeding, vomiting and fever. Classic presentation occurs in older children. In this group, patients with VUR and UTI present abdominal pain as a prime symptom. VUR is an important risk factor for febrile UTI's and renal scarring so it is important to consider it while evaluating patients with abdominal pain and urinary tract infection.
4. Considering the intraabdominal location of the female reproductive tract, different gynecological conditions should be considered as possible aetiologies of abdominal pain. A characteristic example is ovarian masses, especially ovarian torsion. Abdominal pain, nausea, vomiting and palpable abdominal mass is the classical presentation of ovarian torsion, whose diagnosis still remains as one of exclusion. None of the actual imaging studies is definitive in the diagnosis, although ultrasound is useful. The best diagnostic modality in ovarian torsion is laparoscopy; it is also the treatment of choice.
5. Finally, multiple genital anomalies may include abdominal pain as a presenting symptom. Congenital vaginal obstruction as a result of an incomplete canaliza-

tion of the vagina is a diagnosis to consider. An imperforate hymen can result in hydrocolpos, distension of the vagina, and sometimes with distension of the vagina and uterus, known as hydrometrocolpos. These patients are usually diagnosed at newborn age with a palpable lower abdominal mass, urinary tract obstruction, and abdominal pain. If no abdominal mass is present at birth, these patients may remain asymptomatic until adolescence when they present amenorrhea, cyclic abdominal pain and abdominal mass (hematometrocolpos). All these patients should be referred promptly to the specialist.

Indications for Referral

(a) Abdominal pain with urinary tract dilatation at any level.
(b) Urinary tract infection diagnosed during evaluation of a patient with abdominal pain.
(c) Patients with urolithiasis.
(d) Renal mass, ovarian mass and/or pelvic mass, which may present with abdominal pain.
(e) Pelvicureteric junction obstruction ideally at an early age when prenatal diagnosis has been made. In older children it should be considered in patients with pain and/or with abdominal mass.

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Chapter 13
Approach to Abdominal Masses

Anne-Sophie Blais and Armando J. Lorenzo

Key Points
- Pediatric abdominal masses are often discovered incidentally and should prompt immediate evaluation.
- Most abdominal masses originate from the retroperitoneum, with the kidney being one of the most common organ involved.
- The differential diagnosis should be adjusted according to the age of the patient and the characteristics of the mass.
- An abdominal/pelvis ultrasound is commonly the first diagnostic imaging study performed.

Introduction

Most physicians working in pediatrics will encounter at least once in their career a patient with an abdominal mass. Pediatric abdominal masses are often discovered incidentally and should prompt immediate evaluation. Differential diagnosis is extensive and may involve many systems including the gastrointestinal and genitourinary systems. The main challenge for the physician is to differentiate benign from the malignant masses. While most lesions in infants are benign, all children are at risks of developing malignant conditions. Patients with abdominal masses should have a detailed clinical history and a meticulous physical examination. Further investigations should be conducted accordingly in order to clarify the cause of the mass. Imaging studies should follow the ALARA ("as low as reasonably

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achievable") principle, minimizing the use of radiation to the lowest needed in order to secure the diagnosis.

Most abdominal masses originate from the retroperitoneum, with the kidney being the principal organ involved [1]. Of these, the most common renal mass in newborns is hydronephrosis. Renal malignancies are relatively uncommon in pediatrics. Congenital mesoblastic nephroma is the most common neoplasm before 3 months of age; subsequently Wilms tumor increases in frequency and peaks during early childhood. The most common malignant abdominal mass in infants remains the neuroblastoma. It accounts for 8–10% of all childhood cancers [2]. Liver masses account for 5–6% of the pediatric abdominal masses [1]. Hepatoblastoma is the most common primary liver tumor beyond infancy, especially between 1 and 3 years of age. On the other hand, liver hemangioma is common before 6 months of age and typically involutes earlier in life. Table 13.1 reviews the most common causes of abdominal masses in pediatric patients.

History

An organized questionnaire based on the location of the mass and the age of the patient is needed in order to help narrow down the origin and potential nature of the mass. Age is particularly important to adjust the differential diagnosis [3]. Figures 13.1 and 13.2 show the most common causes of abdominal masses in infants, children and adolescents.

Specific questions about the mass can help differentiate a benign from a malignant condition. A mass that has been present for a long time is more likely to be benign. On the other hand, a mass with a rapid growth is more likely to be malignant. Constitutional symptoms -such as anorexia, weight loss, fever, nights sweats- can also be reported and are more common with malignancies. Similarly, an exhaustive review of systems is mandatory due to the extensive differential diagnosis. If abdominal pain is present, the character of the pain, the acuity of onset and radiation should be detailed. New onset of hypertension may be the first sign of an abdominal mass and should prompt further investigations. Loss of appetite, nausea, vomiting, change in bowel patterns can help distinguish genitourinary from gastrointestinal sources. Bowel obstruction, urinary retention or upper urinary tract obstruction can happen with a voluminous abdominal process expanding in the pelvis. These patients will usually display a decrease in bowel movements or urinary output. It is critical to always ask about the presence of hematuria, which would increase the likelihood of a renal malignancy.

Because neuroblastoma can present as paraspinal tumors and abdominal masses can extend to the spine, a neurologic questionnaire may support your diagnosis. The patient should be asked about back pain, changes in sensation or new motor symptoms.

As we get focused in the specific abdominal mass questionnaire, review of the previous medical history and the family history should not be forgotten.

Table 13.1 Differential diagnosis of pediatric abdominal masses

Origin	Disease	Age	Location	Characteristics
Renal	Hydronephrosis	Infant	Flank	Large differential diagnosis: physiologic, ureteropelvic junction obstruction, ureterovesical junction obstruction, vesicoureteral reflux, neurogenic bladder etc.
	Multicystic kidney disease	Infant	Flank	Congenital. Multiple cysts without normal renal parenchyma. The cysts should not be communicating (as opposed to severe hydronephrosis). Second cause of abdominal mass in newborns after hydronephrosis
	Renal cyst	Older	Flank	Multiple cysts or a complex cyst should warrant further investigations
	Angiomyolipoma (AML)	Older	Flank	High risk of bleeding if measures greater than 4 cm. Associated with tuberous sclerosis. Fat poor AML can be difficult to differentiate from other renal tumors on imaging
	Mesoblastic nephroma	Infant	Flank	Mean age at diagnosis is 3.5 months. Most common renal tumor diagnosed on antenatal ultrasound. Usually excellent outcome with radical nephrectomy
	Nephrogenic rests	Children	Flank	Precursor lesions of Wilms tumor. More elliptical or lenticular in shape compared to Wilms tumor. Surveillance is recommended for children older than 12 months old, or patient younger than 12 months old with a previous diagnosis of Wilms
	Wilms (nephroblastoma)	3–5 years	Flank	Six to seven percent of all childhood tumors. Solid mass with or without cystic areas on ultrasound. All patients should undergo CT with contrast or MRI. A tumor rupture should be excluded if the patient present with significant abdominal pain
	Renal cell carcinoma	Older	Flank	Most common renal tumor in the second decade of life. Hematuria is more common than in Wilms. Difficult to distinguish from other renal tumors on imaging
	Multilocular cystic nephroma	Young	Flank	Encapsulated multilocular tumor made of multiple cysts compressing the renal parenchyma. Typically, indistinguishable from a cystic nephroblastoma on imaging
	Rare malignant renal tumors		Flank	Clear cell carcinoma, rhabdoid renal tumor, medullary carcinoma (…)

(continued)

Table 13.1 (continued)

Origin	Disease	Age	Location	Characteristics
Adrenal	Hemorrhage	Infants	Flank	Usually preceded by a trauma (birth, child abuse, blunt trauma) or sepsis. In the early stage of the hematoma, it can appear solid on the ultrasound. With time, the hematoma liquefies and becomes cystic like. The Doppler of the mass should be avascular
	Neuroblastoma	Young	Flank or pelvis	Median age at diagnosis is 19 months. Most primary tumors arise within the abdomen (65%). Associated with many paraneoplastic syndromes: paroxysmal hypertension, palpitations, flushing, headache, acute myoclonic encephalopathy. Diarrhea and hypokalemia may be present if the tumor secretes VIP. Elevated VMA and HVA. On X-ray, the mass may have areas of calcification. The presence of intra-tumoral calcifications and/or vascular encasement can help distinguish from a Wilms tumor
	Pheochromocytoma	Older	Flank	Can present with hypertension, tachycardia, palpitations, heat intolerance, weight loss, headache, congestive heart failure, a decline in school activity (…). The “rule of 10” has been recently challenged for the 10–90% rule
Liver	Hepatoblastoma	First 3 years of life	Right upper quadrant	Most common primary liver tumor in childhood. Can be associated with BWS and familial adenomatous polyposis, but most cases are sporadic. B-hCG and AFP can be elevated
	Hepatocellular carcinoma	Older	Right upper quadrant	Rare entity in children. Mainly in children older than 10 years old. Underlying liver dysfunction is the main predisposing condition. B-hCG and AFP can be elevated
	Hemangioma	Young	Right upper quadrant	Can present with massive hepatomegaly and cardiac failure. Possible involution reported in the first year of life. May be associated with significant morbidity and mortality even if histologically benign

Table 13.1 (continued)

Origin	Disease	Age	Location	Characteristics
Pelvis	Bladder distention	Any age	Pelvis	Large differential diagnosis: Posterior urethral valves, neurogenic bladder, urethral agenesis etc.
	Ovarian teratoma	Older	Pelvis	Most common ovarian neoplasm in childhood. Can be mature (benign) or immature (more aggressive course). The mature type usually has a predominant cystic appearance on imaging
	Others: Hydro/Hematocolpos, anterior myelomeningocele, Sacrococcygeal teratoma (…)			
Intraperitoneal	Urachal cysts, duplication cyst, intussusception, pyloric stenosis, meconium pseudocyst, Omental/mesenteric cyst, Organomegaly, pancreatic pseudocyst, sarcoma, Teratoma, lymphoma (…)			

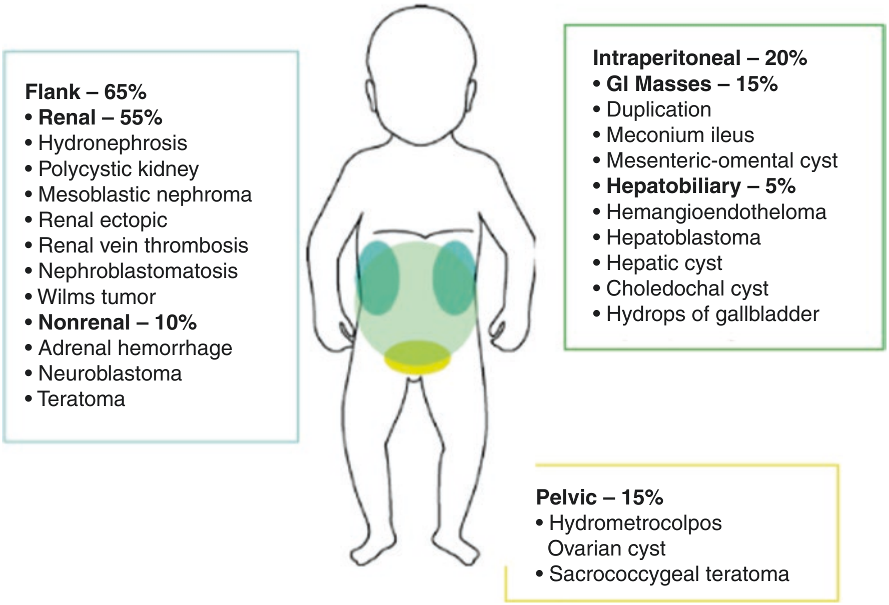

Fig. 13.1 The most common causes of abdominal mass in infants

Prenatal imaging and interventions are important, particularly in newborns and infants. Prematurity is associated with hepatoblastoma. Polyhydramnios may indicate intestinal obstruction. Moreover, a traumatic birth can be suggestive of adrenal hemorrhage. The family should also be questioned on family history of malignancy and syndromes. Beckwith-Wiedemann syndrome (BWS), Denys-

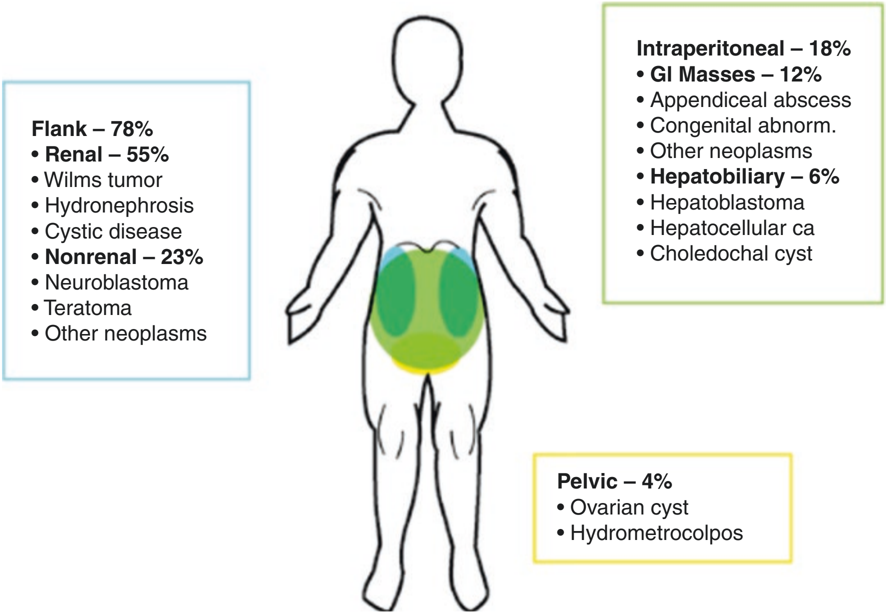

Fig. 13.2 The most common causes of abdominal mass in children and adolescents

Drash and Wilms/aniridia/GU anomalies/mental retardation (WAGR) are well known syndromes associated with Wilms tumors. BWS can also be associated to hepatoblastoma.

Finally, a previous history of trauma or ingestion of foreign bodies should be part of the questionnaire. A foreign body can cause significant enlargement of the stomach and bowel loops and can mimic an abdominal mass. It is usually associated with decreased appetite and signs and symptoms of bowel obstruction.

Physical Examination

A head-to-toe physical examination is mandatory to help determine the origin of the abdominal mass and aid in staging the disease. The patient should be first measured and weighted and the results should be recorded on the growth chart according to their age. Vitals signs should be documented. Hypertension can be seen with neuroblastoma, renal or adrenal masses (Fig. 13.3). Some are endocrinologically active and secrete hormones which can cause signs and symptoms such as tachycardia, flushing or diarrhea. Fever can be caused by a paraneoplastic syndrome, as in lymphomas, or an infection, as hepatitis. A large abdominal mass can cause a restrictive syndrome leading to respiratory embarrassment. Focused inspection of the patient can reveal important clues to the investigation. Asymmetry

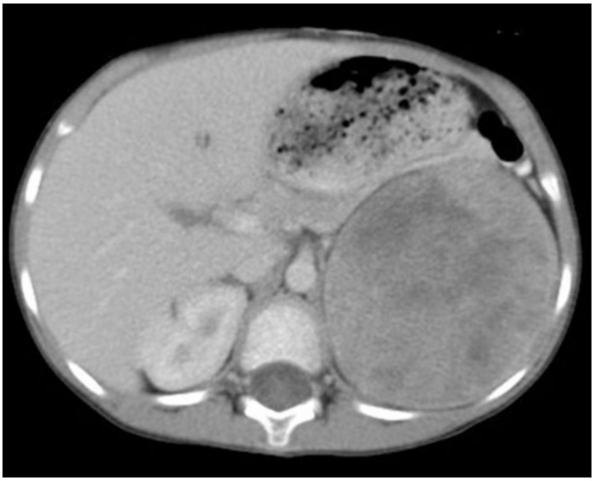

Fig. 13.3 Left adrenal mass causing hypertension due to renin secretion

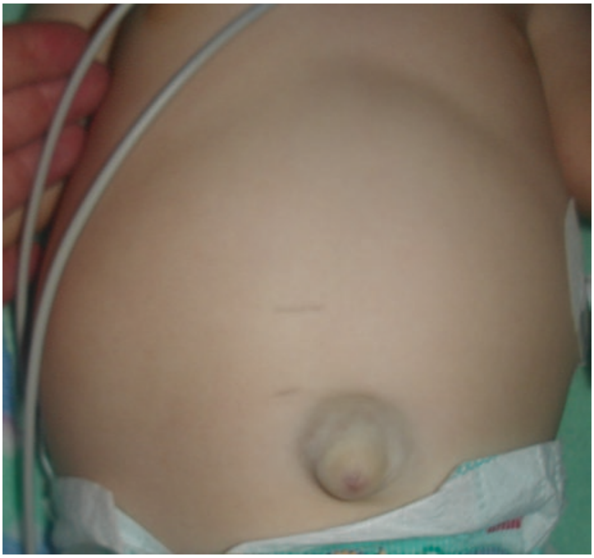

Fig. 13.4 Abdominal bulging due to a large abdominal mass

of the abdomen or significant bulging can be seen in large abdominal mass (Fig. 13.4). A metastatic neuroblastoma should be suspected in the presence of proptosis, periorbital ecchymosis (so called "racoon eyes") or subcutaneous skin nodules (blueberry muffin lesion). Moreover, hemihypertrophy (associated with BWS) (Fig. 13.5) and aniridia (associated with WAGR) (Fig. 13.6) are highly indicative of a Wilms tumor.

An abdominal examination should then be performed. The hands of the examiner should be warmed prior to the exam and the patient should be in a comfortable, quiet environment. The parents can help by distracting a nervous

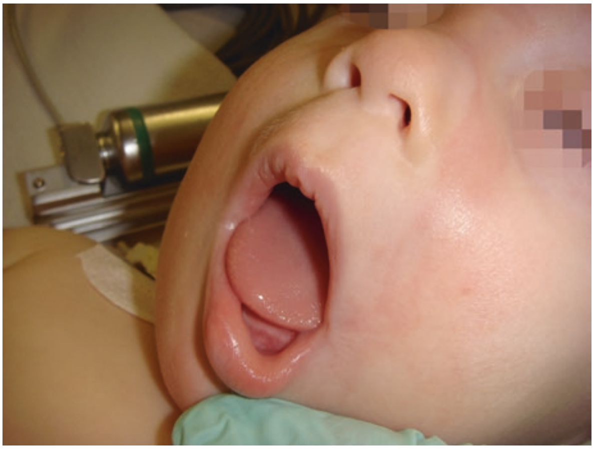

Fig. 13.5 Macroglossia in a patient with BWS

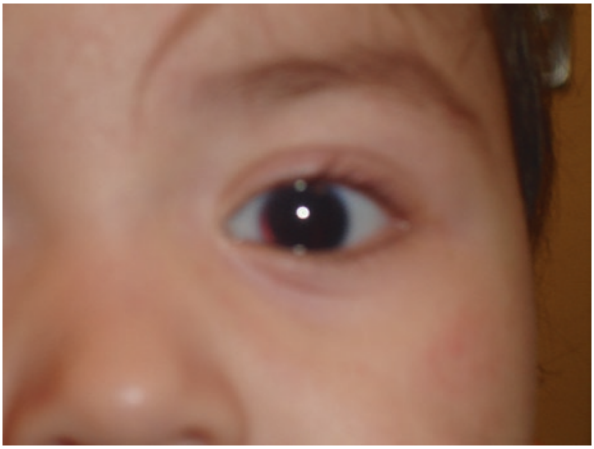

Fig. 13.6 Patient with aniridia

or apprehensive patient. Auscultation of the abdomen should be done to document bowel sounds. Percussion of the abdominal wall can help determine the presence of guarding, ascites and the size of the intraabdominal organs such as liver and spleen. Among others, hepatomegaly can be found with a liver neoplasm or neuroblastoma. The four abdominal quadrants should be palpated. The location, size, tenderness, consistency and mobility of the mass should be documented.

The genital examination can reveal some abnormalities in the case of a Wilms tumor. Ambiguous genitalia, hypospadias and cryptorchidism can be associated with this tumor. Varicoceles can be seen in patients with large abdominal mass (Fig. 13.7). A pelvic abdominal mass can also lead to abnormal genital findings, including processes causing hydrocolpos or hematocolpos.

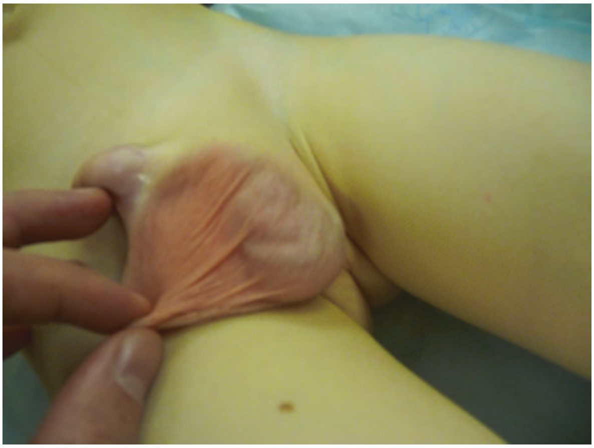

Fig. 13.7 Left varicocele in a patient with abdominal mass

Investigations

Laboratory Tests

Routine bloodwork includes a complete blood count, liver, pancreatic and renal function tests. Anemia, leucopenia or thrombocytopenia can indicate bone marrow infiltration. Electrolytes, uric acid and lactate dehydrogenase abnormalities can be seen in cases with high tumor burden or rapidly growing neoplasms. Hypercalcemia can also happen as part of a paraneoplastic syndrome, as in congenital mesoblastic nephroma, or due to bone metastases. Liver, pancreatic and renal function tests can be altered in liver, pancreatic and renal masses.

More specific blood tests can be performed. Beta-human chorionic gonadotropin and alpha-fetoprotein can be elevated with teratoma, liver and germ cell tumors. Urine analysis should be performed to document hematuria and proteinuria, especially if a tumor of renal origin is suspected. High vanillylmandelic acid and homovanillic acid are usually indicative of a neuroblastoma.

Imaging Studies

An abdominal and pelvic ultrasound is commonly the first imaging study performed when an intraabdominal mass is suspected. In addition to being easy-to-access, inexpensive and safe, it provides information on the location of the mass, the origin, the characteristics (cystic vs solid) and the vascular supply of the mass (Fig. 13.8). If the mass is solid, a computed tomography (CT) or a magnetic resonance imaging

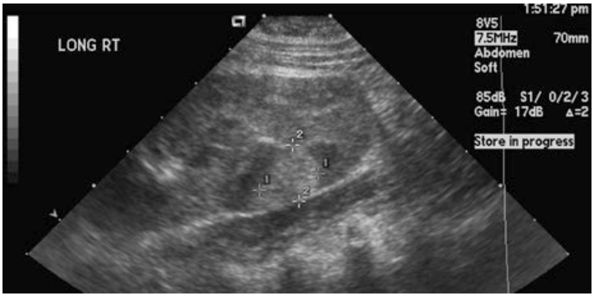

Fig. 13.8 Ultrasound of a right kidney showing a solid upper pole mass

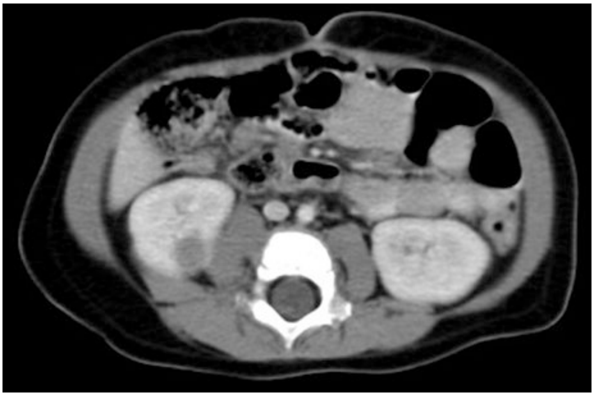

Fig. 13.9 CT imaging of the right kidney mass found on ultrasound

(MRI) is required in most cases to characterize the mass, assess organ involvement and stage the disease (Fig. 13.9).

An abdominal X-ray can sometimes be useful if the mass is thought to be from a gastrointestinal origin. Displacement of bowel loops away from an area of the abdomen is frequently seen with large abdominal masses, and is not specific (Fig. 13.10). Multiple air fluid levels can be seen with bowel obstruction. Severity of constipation can also be documented by the X-Ray. Moreover, calcifications can be seen in neuroblastoma, teratoma, kidney or biliary stone diseases.

In specific cases only, a nuclear scan can be performed. An iodine-131-meta-iodobenzylguanidine (MIBG) can be useful to stage or evaluate the response to therapy of a neuroblastoma. A bone scan can be performed to document bone metastases in case of aggressive tumor that are likely to metastasis to bones or if the patient presents symptoms specific to bone metastases.

If a patient is found to have an abdominal mass that is likely malignant, staging is key to the management plan and should be as thorough as possible. This includes imaging of the chest and -sometimes- the central nervous system in search of metastatic disease. In selected cases, an open or percutaneous biopsy may be indicated. It will lead to either a required surgery, or will obviate unnecessary interventions.

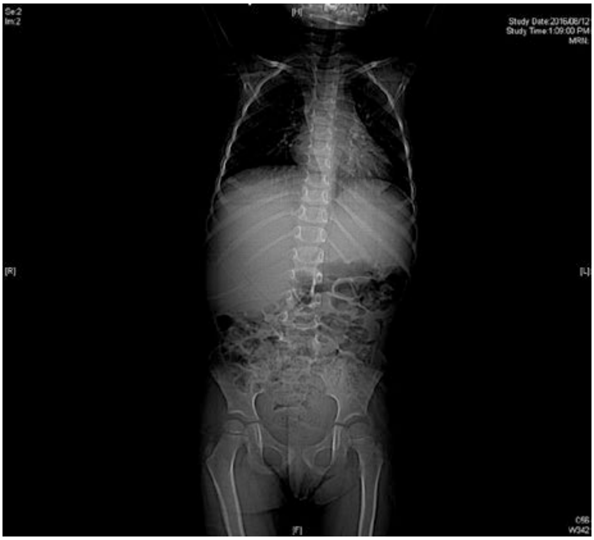

Fig. 13.10 Displacement of bowel loops away from a right upper quadrant abdominal mass

Treatment, prognosis and outcomes of pediatric masses depend on the cause. A stepwise approach, based on a good history and physical examination, will lead to accurate diagnosis and appropriate management.

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Chapter 14
External Angular Dermoid and Pilomatrixoma

Ashish Wakhlu and Nitin Pant

Key Points
- External angular dermoid comprises nearly 61% of all head and neck dermoid cysts.
- They occur typically at the outer corner of the eye as a mass beneath the outer end of the eyebrow.
- Excision is the treatment of choice.

External Angular Dermoid

Introduction

The head and neck region is the commonest site for dermoid cysts in children. External angular dermoid comprises nearly 61% of all head and neck dermoid cysts [1]. They are congenital swellings, but not all are clinically apparent at birth. They occur at the outer corner of the eye typically appearing as a mass beneath the outer end of the eyebrow.

Etiology

Dermoid cyst is a type of choristoma. A choristoma is a benign, congenital proliferation of histologically mature tissue element/s not normally present at the site of occurrence, resulting from either normal tissue migrating to or remaining in an

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abnormal location during embryogenesis. These cysts form at the sites of the lines of embryonic fusion where ectodermal elements either get trapped beneath the skin or fail to disconnect from the developing neural tube.

Clinical Features

Clinically an external angular dermoid presents as a firm, subcutaneous, non-tender swelling under the lateral end of the eyebrow. Despite being attached to the underlying periosteum it is mobile. Occasionally there is a punctum, signaled by deformity of a few eyebrow hairs (Fig. 14.1). Untreated, the swelling grows in size, eventually impinging on opening of the eye. X ray of the skull and/or a non-contrast CT scan is recommended if intracranial protrusion is suspected, although it is rarely required.

Histopathology

These cysts are usually lined by a keratinizing stratified squamous epithelium and its walls commonly contains mature adnexal structures such as terminal hair follicles, sebaceous glands, apocrine glands, and/or eccrine glands [2, 3]. Smooth muscle are sometimes seen within the lining; however, bone and cartilage are uncommon [3]. Cyst generally contains lamellar keratin but infrequently may also contain terminal hair shaft [2, 3]. Despite the findings that the cyst wall shows evidence of chronic granulomatous inflammation, this seldom exhibits clinically [4, 5]. The more chronically the wall is inflamed, the more prone it is for spontaneous or intra-operative rupture [6].

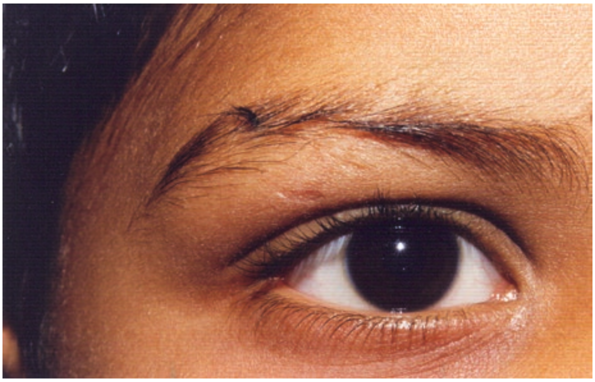

Fig. 14.1 External angular dermoid with punctum

Treatment and Prognosis

The treatment of external angular dermoid is excision, with care to remove the whole lining of the cyst to prevent recurrence. This can usually be accomplished through a skin crease incision. The incision can be given directly over the lesion, above or below the lesion, on the eyebrow or through the natural crease of the upper eyelid. For intracranial extension a craniotomy may be accomplished by lowering a frontal flap. Previously it was thought that intraoperative rupture of cyst lead to post-operative inflammation [7, 8]. In contrast, recent reports advocate that persistent inflammation is more likely after spontaneous rupture than intra-operative rupture [6]. Recurrence is uncommon unless part of the lining has been left behind.

Recently, stealth subcutaneous endoscopic surgery has been used to excise these cysts and is said to improve the cosmetic outcome with the scars being hidden behind the hairline [9, 10]. The three approaches with stealth surgery are the, sub-periosteal, sub-galeal and subcutaneous. Whereas the sub-galeal approach involves increased risk of sensory nerve damage to supraorbital, supratrochlear nerves and frontal branch of facial nerve, the sub-periosteal approach is difficult. The subcutaneous approach is said to avoid these problems.

Pilomatrixoma (Calcifying Epithelioma of Malherbe)

Introduction

It is a benign skin tumor arising from the matrix cells present in the cortex of a hair follicle [11]. As hair follicles are epidermal structures extending into the dermis, pilomatrixoma resides in the dermis or subcutaneous tissue [12].

Epidemiology and Etiology

It is the second most frequently excised pediatric skin mass after dermoid cyst. The lesion occurs notably in patients less than 20 years (peak 8–10 years), frequently in head, neck, upper extremities, front of the trunk (in a hair bearing area) and is mostly solitary [13]. It is more common in white population. The etiology is uncertain and is attributed to a pause in the life cycle of hair follicle secondary to trauma and inflammation [14]. Polyoma virus is also implicated by some [15]. A mutation in exon 3 of the β-catenin gene (CTNNB1) has been identified in most human and murine pilomatricomas. Betacatenin is said to be integral to the terminal differentiation of the hair follicle during embryogenesis. Association with Gardner syndrome, familial adenomatous polyposis, Myotonic muscular dystrophy and Turners syndrome is reported [16–18].

Clinical Features

It presents frequently as a painless, slowly enlarging, oval shaped, subcutaneous nodule, usually mobile and non-tender, but may become painful in cases of inflammation and ulceration. Multiple lesions are seen only in 2–3%cases.The lesion is often tethered to the overlying skin which becomes blue or red blue especially in white population. It feels hard and gritty owing to the calcification within the tumor. Average size of the lesion is 0.5–2 cm. in diameter. Progression takes months to years. With larger lesions thinning of the overlying skin may occur. Notably, a certain preoperative diagnosis based on clinical examination is possible in less than 50% cases [19–22].

Radiological Features

Akin to dermoid cyst, imaging is infrequently used as a supplement to clinical diagnosis. Ultrasound is the most common investigation advised. Features on ultrasound suggesting pilomatrixoma are - A hypoechoic rim seen in (65–82.8%) cases [23–25]. The histopathologic correlate of hypoechoic rim is postulated to be the connective tissue capsule of pilomatrixoma [26]. A second feature on ultrasound is internal reticulations seen in nearly 65.5% cases [27]. A specific but not common feature of pilomatrixoma is the complete calcification of the lesions that generates extensive posterior acoustic shadowing.

These features of a hypoechoic rim, internal reticulations, and calcifications are not seen in dermoid cysts and help in its differentiation from the latter.

Histopathology

Pathological examination shows a keratin core in the center of the lesion with peripheral basophilic (basaloid) cells around this core. These cells are large and pale resembling epidermal keratinocytes and are called as “empty”, “shadow” or “ghost” cells devoid of nuclei, and are pathognomonic of Pilomatrixoma [28]. Moreover there is a fibro-collagenous stroma with multinucleated foreign body type giant cells and small areas of calcium deposition [13].

Treatment and Prognosis

The mainstay of treatment is excision, usually accomplished under general anaesthesia or under local anaesthesia in older children and adults. The tumor is easily removed in its entirety. Recurrence after removal is uncommon (0–3%) [13]. Malignant transformation (carcinoma) is rare and happens in the late middle age.

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Chapter 15
Neck Swellings: Midline Neck Swellings

Rachael Lawrence and Neil Bateman

Key Points

- Thyroglossal duct cysts are the commonest congential abnormality in the neck. They can usually be easily diagnosed clinically.
- The cystic nature of a thyroglossal duct cyst can be confirmed, and the presence of normal thyroid be established, by ultrasound.
- A midline block excision of tissue from the level of the thyroid up to the tongue base, including the middle third of the hyoid bone is the treatment of choice in thyroglossal duct cysts.
- Dermoid cysts can be easily treated by simple surgical excision. If there is any doubt over the diagnosis then the lesion should be treated as a thyroglossal cyst.

Introduction

Neck swellings occurring in children can be usefully divided into those occurring in the midline of the neck, discussed in this chapter, and those situated laterally. While it is uncommon for neck masses to represent sinister pathology, it is important that these lesions are accurately diagnosed and treated by those with appropriate expertise, as the treatment of those children receiving initial inappropriate treatment can

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be very challenging. The differential diagnosis of a midline neck mass in a child includes a thyroglossal duct cyst, midline dermoid cyst, lymph node, lipoma, hemangioma and ranula.

Differential Diagnosis of Midline Neck Lump

Thyroglossal Duct Cyst

Thyroglossal duct cysts are the most common congenital abnormality in the neck. They account for approximately 70% of all congenital neck lesions [1]. The thyroglossal duct is formed by the descent of the thyroid gland in the fetus from the foramen cecum in the tongue base to its familiar position anteriorly in the neck. Under normal circumstances the duct involutes between the eighth and tenth week of gestation. Cysts forming in a persistent duct are clinically known as thyroglossal duct cysts. The thyroglossal duct is closely related to the hyoid bone. This gives rise to the cardinal clinical sign of these lesions, namely elevation on tongue protrusion.

Thyroglossal duct cysts can present at any age although it is very rare for them to be present at birth. A large review of patients with thyroglossal duct cysts showed that although nearly one-third of cases occur in children less than 10 years of age, nearly one-third also occurred in patients in their third decade of life or older [2]. They are almost all in the midline (Fig. 15.1) although they can occur more laterally in the neck. When this happens, it is more commonly on the left. Midline thyroglossal duct cysts are most commonly just inferior to the hyoid at the level of the thyrohyoid membrane. Other locations include the suprahyoid region in 20 to 25% of cases and at the level of the hyoid in 15 to 50% of cases [3].

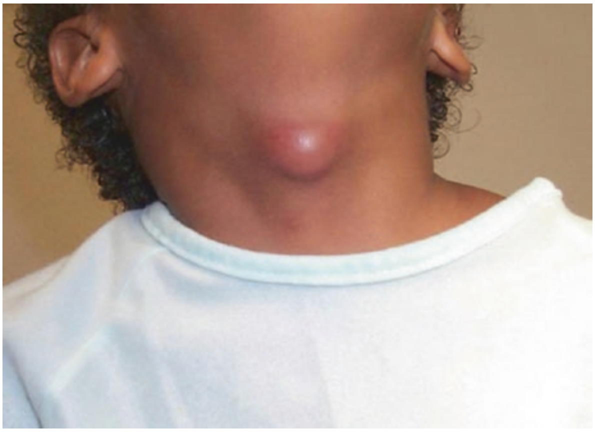

Fig. 15.1 Thyroglossal duct cyst in the midline

Recurrent episodes of infection may occur and, if sufficiently severe and frequent, this may lead to cyst rupture and subsequent sinus formation. It is theoretically possible, although exceptionally rare, that a lesion with the appearance of a thryoglossal duct cyst is, in fact, an incompletely descended thyroid gland and represents the only viable thyroid tissue. There are some case reports of thyroid carcinoma occurring in these lesions, and the incidence is reported to be around 1% [4].

Midline Dermoid Cyst

These are developmental abnormalities caused by abnormalities in midline fusion in the fetus. They appear clinically as midline lesions occurring at any level in the neck. They do not tend to move on tongue protrusion.

Lymph Node

While lymphadenopathy in the neck is most commonly situated laterally, submental nodes may present in the midline.

Thyroid Nodule

Pyramidal lobe nodules or those occurring in the thyroid isthmus may present as a midline lump, although thyroid nodules most commonly occur laterally in the neck.

Ranula

A ranula is a cystic lesion arising from the floor of the mouth. It is essentially a mucous extravasation pseudocyst or a mucocele that can be classified into two types, the much more common simple or intraoral ranula and the plunging, cervical type. A simple ranula is localized to the floor of the mouth. However, with the plunging type, extravasation of mucus occurs beyond the confines of the floor of the mouth and the mucus collection is therefore in the infra-mylohyoid compartment of the neck, with or without a clinically apparent intraoral collection.

A ranula may be congenital but is more often acquired as a result of obstruction to one of the sublingual salivary glands or ducts. It typically presents clinically as a smooth swelling under the tongue with a bluish translucent appearance (Fig. 15.2).

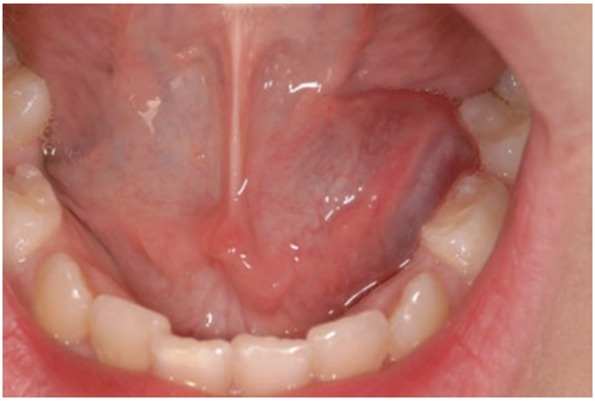

Fig. 15.2 Ranula in floor of the mouth

Investigations

In most children a presumptive clinical diagnosis can be confirmed with ultrasound. This can differentiate between cystic and solid lesions. While ultrasound is a highly user dependent modality, it is often possible to positively identify lymph nodes as well as other common masses such as a lipoma. Ultrasound is the most common initial test for evaluation of a thyroglossal duct cyst and has been shown to have sensitivity and accuracy comparable to CT and MRI [5]. It can also identify the presence of a normal thyroid gland. Occasionally imaging modalities such as MRI are called for, although these are more commonly used in larger, more complex neck masses. Fine needle aspiration, while in adults the investigation of choice for many neck lumps, is very rarely helpful in investigating neck lumps in children.

Treatment

Thyroglossal Duct Cyst

The treatment is surgical, with excision of the cyst and any associated tract, including the mid-third of the hyoid bone. The classically taught operation where the cyst and tract are identified and followed to the tongue base has been largely superseded by a midline block dissection of tissue from the thyroid isthmus up to the foramen cecum. The deep limit of the dissection is the trachea and laryngeal cartilages. The rationale behind this is that the thyroglossal duct is a complex branching structure and this technique is less likely to leave portions of duct remaining, which carry the risk of recurrence.

Midline Dermoid Cyst

Simple excision is curative. At surgery, they are superficial and have a characteristic yellow/white colour. If there is any doubt then after excision the cyst can be bivalved. If contains sebaceous material then the diagnosis of a dermoid is confirm. If not then it should be assumed to be a thryoglossal duct cyst and treated appropriately.

Lymph Nodes

These can be managed as any other lymph node in the neck (see Chap. 18).

Thyroid Nodule

In comparison with thyroid neoplasms in adults, the pediatric population exhibits differences in pathophysiology, clinical presentation and long-term outcomes. Hence, The American Thyroid Association recognized that therapy for an adult might not be appropriate for a child who is at low risk of death but at a higher risk for long-term harm from overly aggressive treatment. Management guidelines for children and adolescents <18 years with thyroid nodules and differentiated thyroid cancer were therefore developed and published by this association in 2015 [6].

Ranula

Simple aspiration or drainage of a ranula results in a high recurrence rate. Marsupialization of a plunging ranula is reported to be inadequate with a recurrence rate of 80% [7]. Both intraoral and plunging ranulas should be excised, together with the cyst wall and the sublingual gland as excision of the gland results in a low rate of recurrence [8, 9]. Excision carries a risk of damage to structures in the floor of mouth including the hypoglossal nerves.

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Chapter 16

Neck Swellings: Lateral Neck Masses

Neil Bateman

Key Points

- Lymphadenopathy in children is the commonest cause of laterally placed neck lumps.
- The majority of children with a lymph node in the neck have self-limiting viral induced reactive lymphadenopathy.
- Referral and/or biopsy is indicated where there is persistent adenopathy greater than 2 cm in diameter, systemic symptoms, nodes in multiple sites or where the lump has a sinister clinical or ultrasound appearance.
- Cancer is a common cause of death in children. Where there is any doubt about the cause of a neck lump then an urgent biopsy should be considered.
- Haemangiomas are benign vascular tumours and are an entirely distinct clinical entity from vascular malformations and hence require different treatment strategies.

Introduction

The majority of laterally placed neck lumps in children are of lymph node origin. Cervical lymphadenopathy is very common in children and in the vast majority of children will be self limiting and benign. Clinicians must therefore be aware of the potential causes of lymph node enlargement in children and the indicators of serious pathology.

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Differential Diagnosis of Lateral Neck Lumps

Lymph Nodes

The causes of lymphadenopathy in the neck are;

Infective

Viral Upper respiratory viral infections may cause lymphadenopathy, which is usually self-limiting. Other common childhood viral infections such as measles, mumps, and rubella can also cause lymphadenopathy. Infectious mononucleosis, caused by Epstein Barr virus infection, can cause massive lymphadenopathy, often accompanied by tonsillitis, fever, lethargy, and malaise. The diagnosis can often be confirmed with a serological test (such as the Monospot or Paul Bunnell) and there are often atypical lymphocytes on the blood film.

Human Immunodeficiency Virus (HIV) may present with persistent generalized lymphadenopathy. The transmission in children is usually vertical from mother to child. They may be a history of recurrent or opportunistic infections. This is a diagnosis that should be considered in any patient with an unknown cause of lymphadenopathy.

Bacterial Acute lymphadenitis may occur with infection with pyogenic organisms, most commonly streptococci and staphylococci. Occasionally gram-negative organisms may be responsible. If untreated this may progress to an acute inflammatory mass and/or suppuration to form an abscess (Fig. 16.1). Often these are relatively superficial and require an external incision and drainage. Infection within the deep neck spaces can present more of a management challenge and carries a risk of airway obstruction or progression to the mediastinum [1].

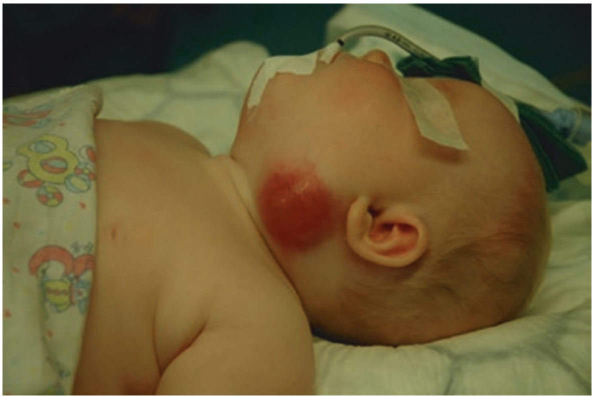

Fig. 16.1 A superficial neck abscess in an infant

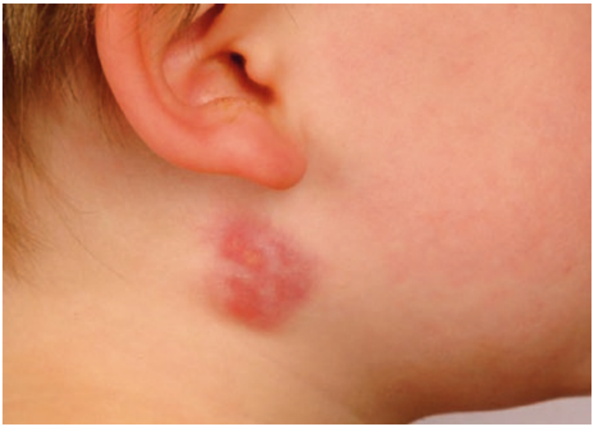

Fig. 16.2 Non-tuberculous mycobacterial infection in the parotid gland showing the characteristic skin discoloration

Mycobacterial Infection Tuberculosis (TB) presenting in the neck is rare in UK practice. More common is non-tuberculous mycobacterial infection, sometimes referred to as atypical mycobacterial infection or incorrectly as atypical TB. These organism cause infection via an oral route and tend to infect those lymph nodes draining the oral cavity (i.e. submandibular and intraparotid nodes most commonly). They most commonly present as a painless mass in an otherwise entirely healthy child. If left untreated the skin over the lump develops a violaceous discoloration and given more time eventually breaks down to form a discharging sinus (Fig. 16.2). If left untreated the natural history of the condition is to spontaneous resolution over the course of several months. Treatment of this condition is either conservative, medical (and a variety of antibiotics have been employed including clarithromycin, azithromycin, ciprofloxacin as well as more traditional antituberculous drugs) or surgery [2]. The resistance of mycobacteria to antibiotic treatment means that, if medical treatment is planned, antibiotic treatment often requires several weeks or months of treatment.

Other bacterial infection, which can present with lymphadenopathy include brucellosis, cat scratch disease, actinomycosis, and syphilis.

Inflammatory

Kawasaki disease is an inflammatory, non-infective condition characterized by a medium vessel vasculitis that presents with cervical lymphadenopathy, pyrexia, desquamation of the hands and feet, erythema of the oral cavity and conjunctivitis. It is an important diagnosis to consider as it carries a potential mortality of 1–2% due to coronary arterial lesions and associated cardiac complications. The treatment is with the administration of intravenous immunoglobulin.

Neoplastic

Fortunately head and neck malignancy is rare in children. However, in 2016 cancer accounted for 20.6% of all children aged 1–15 years [3], and a significant proportion of pediatric cancers involve the head and neck at some stage. Early diagnosis and appropriate management in a specialist center is of vital importance to a good outcome.

Hodgkin's disease occurs most commonly in teenagers and young adults and characteristically affects lymph nodes rather than extra-nodal sites. The typical history is progressive enlargement of cervical lymph nodes. There may be systemic symptoms such as anorexia, weight loss and night sweats. Hepatosplenomegaly indicates more advanced spread of the disease. Diagnosis is by lymph node biopsy and treatment by chemotherapy and/or radiotherapy. Non-Hodgkin's Lymphoma is a heterogeneous group of malignancies distinguished pathologically from Hodgkin's disease by the absence of Reed Sternberg cells. Extra-nodal sites (e.g. tonsil, adenoids) may be affected in the head and neck rather than lymph nodes. A younger age group (2–12 years) is more typically affected. Diagnosis is by a biopsy and treatment modalities similar to that of Hodgkin's disease. Other malignancies presenting as neck lumps include rhabdomyosarcoma, thyroid malignancy and salivary gland tumors. Congenital teratomata may present as large masses in the head and neck.

Vascular Anomalies

In 2014, The International Society For The Study of Vascular Anomalies (ISSVA) developed a classification system for vascular anomalies. Put simply, vascular anomalies are separated into two main groups; vascular tumors and vascular malformations.

With respect to vascular tumors, these are further classified as benign, locally aggressive or borderline, and malignant. Benign vascular tumors consist of various types of hemangioma. Hemangiomas should be clearly distinguished from vascular malformations. Hemangiomas are benign neoplasms of endothelium and grow rapidly in the first few weeks of life, often having been undetectable at birth. They commonly affect the parotid area and can appear alarming to parents and doctors alike. They are known to respond to systemic steroids and propranolol treatment and are also amenable to surgical excision. However, they will if left alone, involute spontaneously so treatment is only really required for lesions causing a functional problem (such as swallowing or airway problems, or in facial lesions where there is closure of one eye). Locally aggressive or borderline and malignant vascular tumors are less common and are outside the scope of discussion for this chapter [4].

Vascular malformations are present at birth and are a developmental abnormality. They include capillary, lymphatic, venous and arteriovenous malformations.

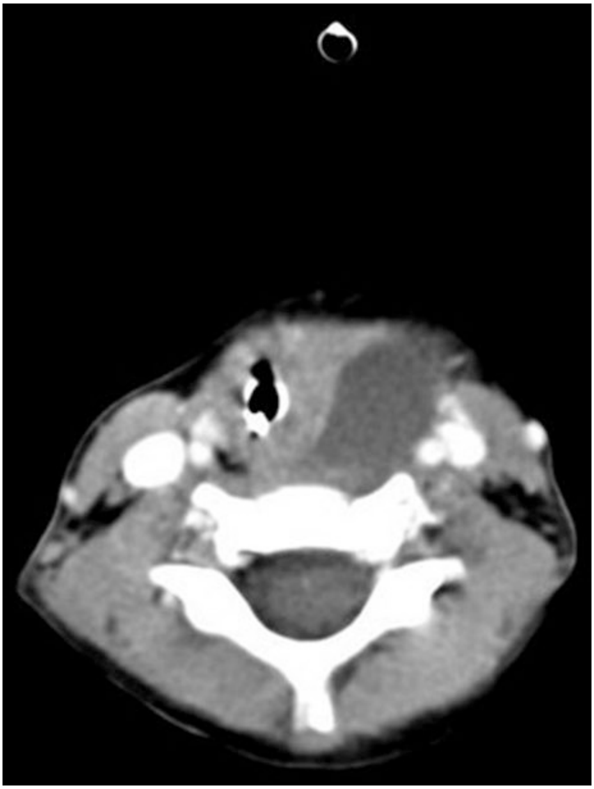

Fig. 16.3 CT scan showing multiloculated lymphatic malformation in the parapharyngeal space extending inferiorly on the left side of the neck. There was airway obstruction and the child presented with snoring

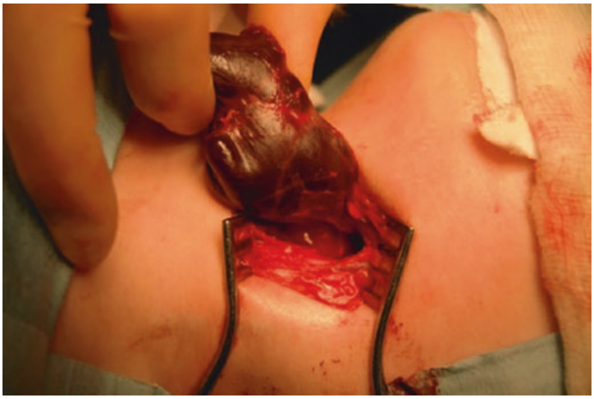

Fig. 16.4 The lesion in Fig. 16.3 at the time of surgical excision

Lymphatic malformations, otherwise known as cystic hygroma or lymphangioma (Fig. 16.3), do not proliferate and do not resolve spontaneously. Treatment is either conservative, surgical (where the functional or cosmetic problem outweighs the implications of surgery), or, in the case of lymphatic malformations sclerotherapy may be considered (Fig. 16.4).

Investigations

Investigations for lymphadenopathy are aimed at determining the likelihood of there being any sinister underlying pathology in order to identify those children who should undergo excision biopsy. Lymph nodes in the neck are a very common problem and many children would undergo unnecessary surgery if they were all biopsied. Ultrasound is a very useful modality to identify numbers and size of nodes as well as abnormalities in nodal morphology.

The following investigations and their significance should be considered;

- CXR may indicate mediastinal lymphadenopathy in children with lymphoma.
- FBC may be abnormal in patients with hematological malignancy, although a normal result does not exclude sinister pathology.
- Serology including Epstein-Barr Virus (EBV), Cytomegalovirus, Toxoplasma and Bartonella may be helpful in establishing an infective diagnosis.
- Fine needle aspiration is uncommonly performed in children. It is less acceptable for patients and parents than in adults and FNAC has limited usefulness in the diagnosis of lymphoma.

Excision biopsy is, in general, indicated in those children:

- Who have multiple nodes in many areas of the neck, especially those with supraclavicular nodes (which have an increased likelihood of sinister pathology)
- Who have a fixed node or one with obvious skin involvement
- Who have persistent nodes of greater than 2 cm diameter
- Who have abnormalities on CXR or FBC
- Who have unexplained systemic symptoms (e.g., anorexia, weight loss, night sweats, etc.)
- Who have abnormal nodal morphology identified on ultrasound.

An evidence based algorithm for the management of pediatric cervical lymphadenopathy has recently been published [5].

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Chapter 17
Neck Swelling/Lumps: Torticollis

Simone Schaefer

Key Points
- Torticollis is a congenital or acquired condition seen in childhood and characterized by a lateral head tilt and a chin rotation towards the opposite side of the tilt.
- Congenital muscular torticollis is the most common aetiology in infancy and often resolves by the age of 1 year.
- Timely referral to a paediatric physiotherapist can prevent secondary sequalae such as plagiocephaly, facial asymmetry and functional delay.
- Non muscular torticollis can be associated with vertebral or neurological abnormalities and requires referral to a secondary service.

Introduction

Torticollis is a condition characterized by a lateral head tilt (rotation of the head and neck to the affected side) and a chin rotation towards the opposite side of the tilt. This can be congenital or acquired and can be seen from infancy throughout adolescence. Depending on the age at presentation, the differential diagnosis varies. The incidence of congenital torticollis ranges from 0.3% to 2% of new-borns and it may occur slightly more frequently in males. For acquired torticollis, the incidence is depending on the underlying aetiology. This chapter will define the two types of torticollis and discuss the evaluation and management of both, including reasons for referral.

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Aetiology and Evaluation of Torticollis

Congenital Muscular Torticollis

Congenital muscular torticollis (CMT) is the most common aetiology of torticollis in new-borns and infants. It is associated with unilateral shortening or fibrosis of the sternocleidomastoid (SCM) muscle and it may be accompanied by cranial deformation, hip dysplasia, brachial plexus injury and distal extremity deformities. Risk factors identified for CMT are plagiocephaly, breech delivery, forceps delivery and twin birth. More controversial aetiological factors comprise of an ischaemic event, venous occlusion, genetic factors, constitutional growth arrest, infective myositis, neurogenesis and intrauterine compartment syndrome.

CMT is traditionally categorized in three types:

1. SCM mass CMT: most common form, also known as fibromatosis colli; consists of torticollis with a palpable mass in the SCM and passive range of movement (ROM) restrictions.
2. Muscular CMT: consists of SCM tightness and passive ROM limitations, but no palpable mass.
3. Postural CMT: consists of the postural preference of the infant, without muscle tightness or ROM restrictions.

Initial evaluation comprises of physical examination of the neck followed by a visual and neurological screen. Depending on the findings, an ultrasound scan might be helpful to identify the cause and differentiate from other pathologies in the neck. MRI for CMT is no longer considered necessary or cost-effective.

Non-muscular Torticollis

Non-muscular torticollis (NMT) or acquired torticollis has a wide variety of aetiological factors, with over 80 different entities described in literature. The most common causes of NMT include Klippel-Feil syndrome (fusion of two or more cervical vertebrae) and congenital scoliosis (30.2% of all NMT), ocular disorders (22.6%), obstetrical palsies (17%) and central nervous system abnormalities (11.3%). Torticollis in the older child is more often a result of trauma or inflammation (Grisel's syndrome), where cervical dystonia might be the underlying cause in older adolescents.

Work-up of non-muscular causes of torticollis can be performed based on the algorithm described by Ballock et al. (Fig. 17.1). After CMT is excluded (based on early presentation and palpable mass in SCM), further evaluation should be focussed on ruling out/identifying any of the above-mentioned aetiologies including history of birth trauma, eye examination, neurological examination (possibly including MRI) and bony abnormalities with the help of cervical spine X-ray. Treatment of NMT will depend on the aetiology.

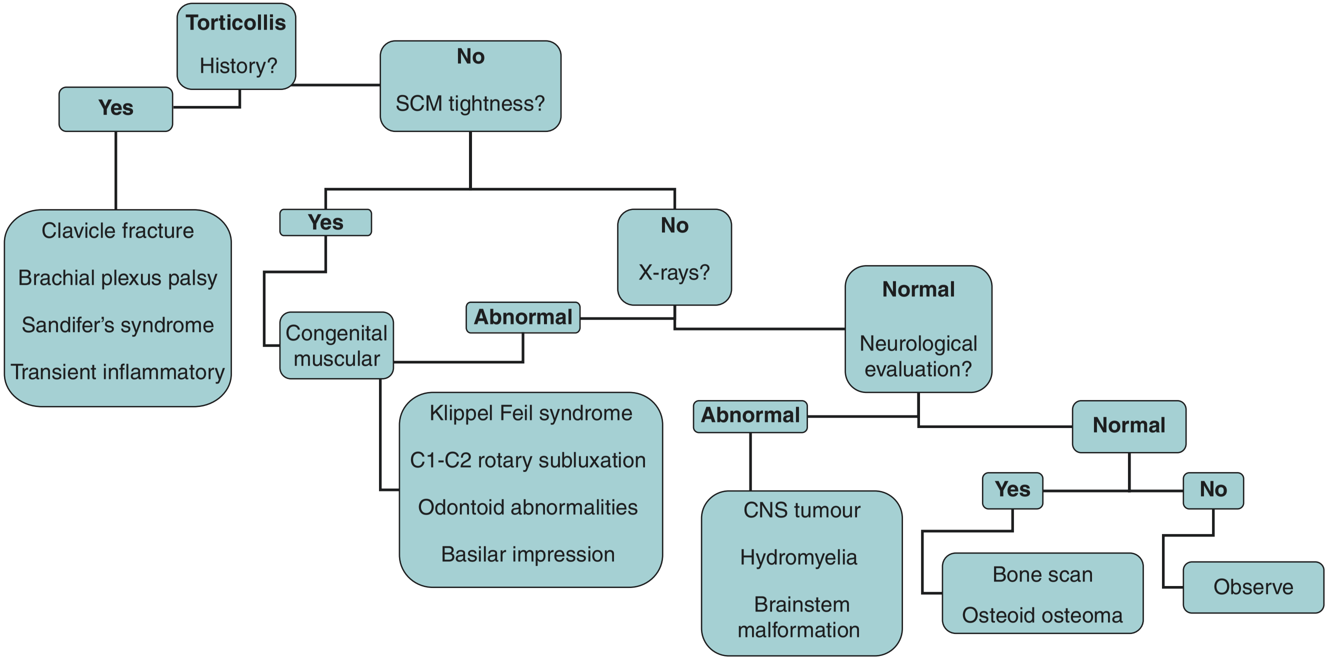

Fig. 17.1 Algorithm for the management of torticollis (adapted from Ballock et al. 1996)

Treatment of CTM

Conservative Treatment

There is strong evidence that early start of treatment results in the best outcomes and translates to prevention of secondary sequalae, such as plagiocephaly, facial asymmetry and functional delay. An infant diagnosed with CMT should therefore be referred to a physiotherapist for further management. This can consist of handling and positioning advice for mild cases as well as manual stretching for moderate to severe cases. A soft collar or brace can be used in conjunction with physiotherapy, mainly for children aged 4–6 months. Most CMT will resolve within 2–6 months of age.

Surgical Treatment

Over the past years, Botox (botulinium toxin type A) injection has been shown to be an effective intermediate method of therapy for more resistant cases of CMT. For more severe or persistent cases, a variety of surgical strategies exists, which can be applied based on patient factors and surgeon's preference. Examples are myotomy, uni- or bipolar release of the SCM, z-plasty or myeloplasty and lengthening of the SCM.

Reasons for Referral

1. Although mild forms of CMT often resolve spontaneously, early intervention with manual stretching has shown additional benefits. Health professionals are therefore advised to refer an infant with CMT to a paediatric physiotherapist.
2. Patients with CMT should be referred to a secondary care centre if they are not making the expected progress with physiotherapy (no improvement after 4–6 weeks of intense initial treatment or moderate improvement after 6 months of treatment), initial presentation after 7 months of age with a SCM mass, or if the infant is older than 12 months on initial presentation with facial asymmetry and/or 10–15° difference persists between left and right for any motion.
3. If NMT is suspected, the underlying cause should be investigated and an appropriate referral should be made to either orthopaedics, neurology, neurosurgery or ophthalmology.

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Chapter 18
Umbilical Disorders

Spencer W. Beasley

Key Points
- Umbilical hernias are common in small children, occurring in about 10% of infants after separation of the umbilical cord, but the vast majority of them involute spontaneously.
- Umbilical hernias virtually never ulcerate, rupture, strangulate or give pain.
- Umbilical hernias only need to be repaired if they are still present beyond the age of 3–4 years: this is done as a day case under a short general anesthetic.
- A moist lump at the umbilicus is usually an umbilical granuloma or ectopic bowel mucosa, and can be treated with topical application of silver nitrate.
- Discharge of air, or ileal content in the presence of a small opening suggests a vitellointestinal tract; and discharge of urine from an opening at the umbilicus indicates a patent urachus.
- Exomphalos and gastroschisis are normally diagnosed on antenatal ultrasonography, and are very obvious at birth.

Introduction

The umbilicus can be the site of a variety of abnormalities, most of which are minor. The most common is the umbilical hernia, and about 90% of these involute spontaneously, usually in the first few years of life. In contrast to inguinal hernias, strangulation of an umbilical hernia is extremely uncommon.

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Common Umbilical Conditions

1. Umbilical hernia: Umbilical hernia presents as a skin-covered swelling at the umbilicus, first seen after separation of the cord (Fig. 18.1). It increases in size with increasing abdominal pressure, (e.g., with crying, straining) but reduces when the infant is relaxed.
2. Umbilical granuloma: this appears as a fleshy pink protuberance which is moist, causing yellow staining of the overlying clothing. It first becomes apparent after separation of the umbilical cord, and has no sinus opening (Fig. 18.2a).
3. Ectopic bowel mucosa at the umbilicus: this has an appearance similar to that of an umbilical granuloma but tends to be more "cherry red" and may bleed more readily.
4. Patent urachus: there is an opening at the umbilicus through which urine passes (because there is a connection with the bladder). A urachal remnant may also

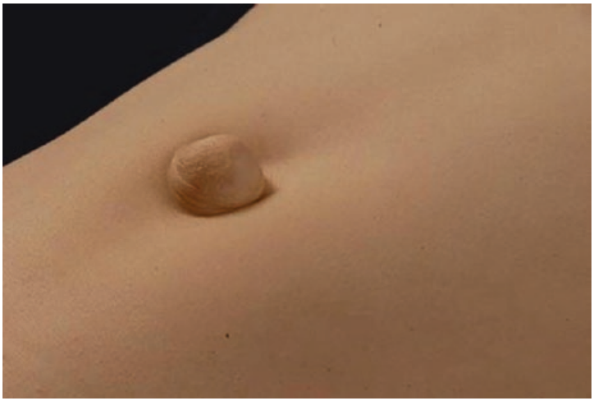

Fig. 18.1 Typical appearance of an umbilical hernia

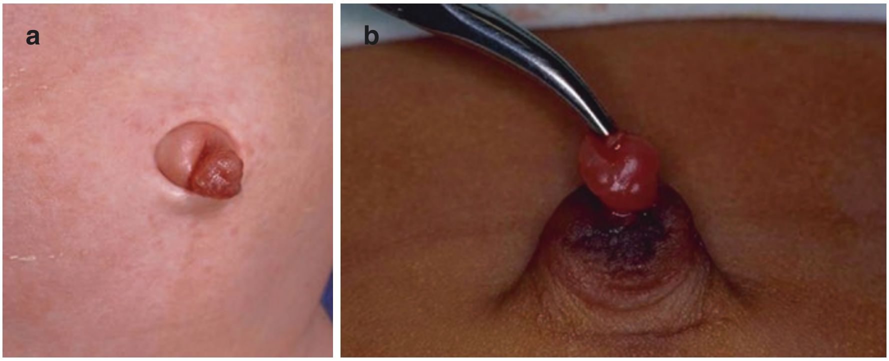

Fig. 18.2 (**a**) An umbilical granuloma presents as a moist and fleshy protuberance at the umbilicus that has no sinus opening. (**b**) An umbilical granuloma often has a "stalk." Here the granuloma is being held up by forceps to show its relatively narrow base. This is where the silver nitrate stick should be applied

present as a blind ending sinus—which may intermittently discharge—or as a urachal abscess inferior to the umbilicus. An abscess will present with midline lower abdominal pain and tenderness, and a mass may be palpable between the umbilicus and bladder. An ultrasound scan will confirm the diagnosis.

5. Patent vitellointestinal tract: there is an open communication between the ileum and the umbilicus. Gas and fluid ileal contents can pass through it, appearing from a small opening on the umbilicus (Fig. 18.3). There are a number of variants of this condition, including a sinus, Meckel's diverticulum and Meckel's band.
6. Exomphalos (or omphalocele): the umbilicus is widely open, and bowel and liver protrude through the broad umbilical cicatrix, but are contained by a sac consisting of peritoneum and an amniotic layer. Often, the infant has associated abnormalities such as cardiac and renal defects, or Beckwith-Wiedemann Syndrome (BWS). BWS should be suspected if the infant is large, has macroglossia, grooved earlobes and a relatively small exomphalos (Fig. 18.4). Early recognition of this condition is important to prevent the profound hypoglycaemia that occurs in the first few days of life.

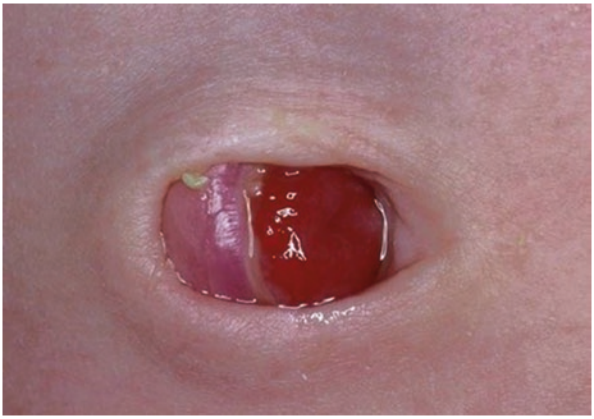

Fig. 18.3 A patent vitello-intestinal tract has an opening through which fluid ileal contents and air can leak. Often this causes redness and excoriation of the skin surrounding the umbilicus

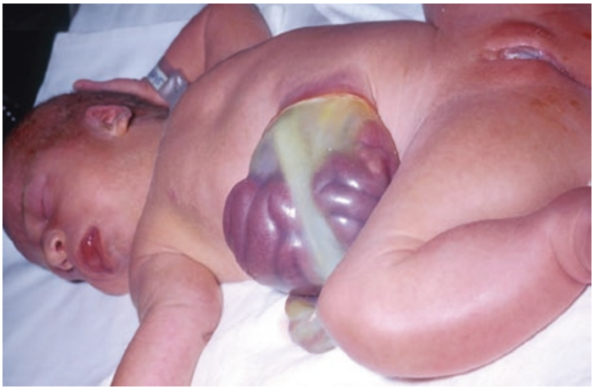

Fig. 18.4 An infant with exomphalos who also has Beckwith-Wiedemann Syndrome. Note the baby is large, has macroglossia and abnormal facies. These infants are at risk of severe hypoglycaemia

7. Gastroschisis: there is a relatively small defect immediately to the right of the umbilicus through which loops of mostly small bowel (and sometimes the gonads) protrude antenatally, without any evidence of a sac (thus distinguishing it from exomphalos). Like exomphalos it is typically detected on antenatal ultrasonography. The bowel is often oedematous and thickened, and there may be an associated bowel atresia. Perforation of the extruded gut may occur.
8. Omphalitis: this is a neonatal infection emanating from the umbilicus. It becomes apparent when streaks of erythema are observed radiating from the umbilicus.

Treatment of Conditions of the Umbilicus

1. Umbilical hernia: Those hernias that do not resolve spontaneously are repaired after 3 years of age as a day case under a short general anesthetic. The skin incision is made within the umbilicus. Recurrence is extremely unusual.
2. An umbilical granuloma and ectopic bowel mucosa are treated either with topical application of silver nitrate to the base of the stalk, or by ligation of the (often narrow) base with a suture tied tightly. Care should be taken to ensure that there is no sinus opening, as this may indicate an underlying vitellointestinal tract or urachus.
3. Where a patent urachus is suspected, imaging with a sinugram or ultrasonography may reveal the extent of the abnormality. Treatment involves surgical excision. This can normally be performed through the umbilicus itself with the dissection remaining extra-peritoneal. A urachal cyst can be dealt with either by an open or laparoscopic approach.
4. Vitellointestinal tract: this also can be confirmed by a sinugram, although a clearly patent vitellointestinal tract may require no further investigation. Treatment is by surgical excision of the entire tract. This can be performed through the umbilical cicatrix.
5. Exomphalos (or omphalocele): this is often first diagnosed on antenatal ultrasonography. Its characteristic appearance makes it easily diagnosed at birth. Associated abnormalities (e.g., renal, cardiac, and chromosomal) should be excluded. In BWS hypoglycaemia must be prevented by glucose infusion. Emergency management involves prevention of excessive heat loss (wrap the torso including the defect in plastic film and place the infant in a warm humidicrib), insertion of an intravenous line, and nasogastric de-compression. These children are then treated in a tertiary neonatal unit with access to specialist paediatric surgical expertise. Surgery returns the bowel contents and liver to the abdomen and the defect at the umbilical ring is closed. For extremely large defects, preliminary non-surgical treatment by external compression, but keeping the sac intact, may be required.
6. Gastroschisis: this is a surgical emergency. There are significant advantages in getting the bowel returned to the abdominal cavity within hours of birth, before it becomes more oedematous and congested. Immediate management at birth involves protection of the bowel by wrapping it in plastic film, preventing heat

loss, and insertion of a nasogastric tube with regular suction to keep the bowel empty of air and fluid. Surgery involves returning the bowel to the abdominal cavity as soon after birth as possible, with closure of the defect. Sometimes, the volume of the extra-abdominal bowel and the comparatively small abdominal cavity into which it must be placed, means that a temporary silo has to be fashioned.

Indications for Referral

1. An umbilical hernia that has not resolved by the age of 3–4 years.
2. A presumed umbilical granuloma/ectopic bowel mucosa unresponsive to ligation or topical application with Silver Nitrate.
3. Suspicion of a patent vitellointestinal tract or urachus (both conditions will have a sinus opening evident at the umbilicus).
4. All exomphalos and gastroschisis infants: antenatal diagnosis allows transfer to a pediatric surgical centre prior to birth; failing that, the newborn infant is transported to the tertiary centre as soon after birth as possible, with care taken to support the defect, preventing hypothermia, and providing nasogastric suction and intravenous fluid resuscitation.

Epigastric Hernia

Introduction

An epigastric hernia becomes apparent as a small midline lump, usually midway between the xiphisternum and umbilicus (Fig. 18.5). The swelling is often less than a centimeter in diameter. The bulge is due to extra-peritoneal fat protruding through a small defect in the linea alba.

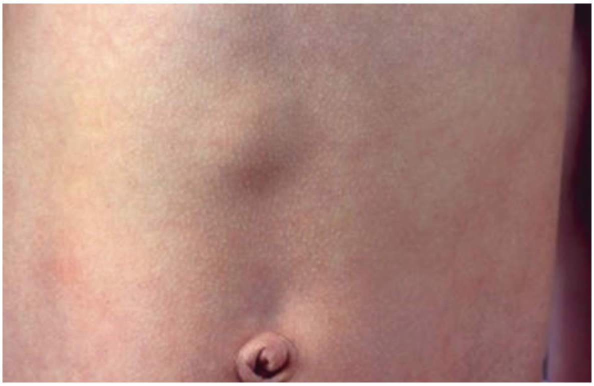

Fig. 18.5 An epigastric hernia appears as a midline lump usually midway between the xiphisternum and umbilicus

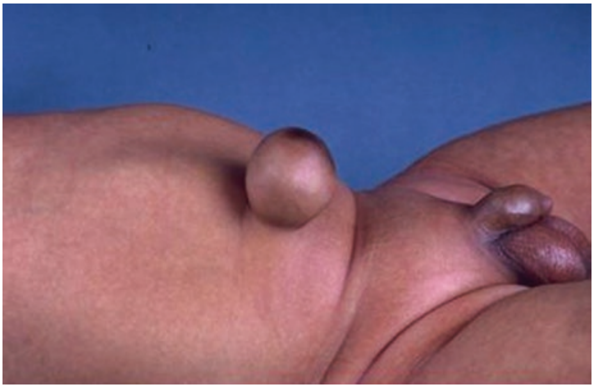

Fig. 18.6 Divarication of the rectus abdominus muscles in association with an umbilical hernia

The parents typically notice a small non-tender swelling in the midline or immediately adjacent to it. It may be more prominent or cause discomfort after meals and exercise.

It is a relatively harmless condition, but if it causes concern or discomfort, it can be repaired under a short general anesthetic as a day case with excellent results.

Divarication of the Rectus Abdominus Muscle

This is best considered a variant of normal, rather than an abnormality. Often, in infants and small children, there is a longitudinal bulge between the xiphisternum and the umbilicus that is most obvious when the abdominal muscles (rectus abdominus) are contracted. This can be demonstrated by getting the child to lift his/her head up while lying supine (Fig. 18.6).

It is of no significance, other than it often causes concern to parents or a medical practitioner unfamiliar with the condition.

No treatment is required. It has no long term sequelae. It never causes discomfort.

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Chapter 19
Surgical Aspects of Abdominal Pain

Jonathan Goring and Govind V. S. Murthi

Key Points
- Abdominal pain in children is common and usually follows a benign and self-limiting course but can herald significant surgical pathology.
- Analgesia should be used in children with abdominal pain and does not "mask" symptoms.
- Repeated examinations and/or careful safety net advice should be considered so that an initial diagnosis can evolve to be confirmed or resolution of symptoms occur to avoid missing a potentially serious diagnosis.
- Remember to think broadly about where the source of the pain is—the abdomen is sometimes not the primary site of pathology; it can originate in the chest, scrotum, groin, back or spine.

Introduction

Abdominal pain is common in children with studies showing that up to 40% of children experience at least one episode every week. This is reflected in the frequency with which both primary and secondary care practitioners encounter children presenting with this problem. The majority of these children have a benign self-limiting problem and no surgical intervention is required. However abdominal pain can be the presenting feature of significant pathology that warrants prompt surgical management. Deciding which patients do or do not need referral to a surgical unit can be difficult. This chapter aims to help the primary care practitioner

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make this decision with a guide to the most common diagnoses, how to differentiate between them and how to avoid pitfalls in managing these difficult clinical scenarios.

General Approach to a Child with Abdominal Pain

Whilst the list of potential causes of a child's abdominal pain is wide and varied, involving potentially many systems of the body (ENT, Respiratory, Metabolic etc.) a thorough, systematic history and examination should exclude these other potential causes of abdominal pain. The essence of most consultations, boils down to differentiating non-specific abdominal pain (NSAP) from acute appendicitis.

History

Taking a history from a paediatric patient can very often be as equally informative and rewarding as that from an adult patient however at times the combination of pain, a new environment and a strange doctor unknown to the child can lead to difficulties in elucidating the key features of the abdominal pain. Parents and carers are key here to gain information, however don't forget to question the child themselves if feasible. Bear in mind that children will sometimes say "yes" to questions to try to please their doctor, or deny pain as they are scared what may happen to them if they admit to it, so question openly, sympathetically and use language appropriate to the child's age.

Examination

Depending on the child's age, the clinician may have to take a flexible or inventive approach to examining their patient. Always keep the child's primary carer in view during the examination and never start with palpating the tender area—you will lose their trust immediately and you may not get another go! Distraction is key to a successful examination as almost all children will voluntarily "guard" their abdominal musculature if they think you are going to hurt them. Therefore, a friendly chat with the child about their favourite football team, school friend etc. whilst palpating the abdomen can really help you decide if the abdomen is truly soft or not. Unfortunately, eliciting tenderness is a key part of the examination, however this should be minimised wherever possible. Once again, some children will not overtly tell their doctor if they are in pain and it is often a grimace or change of breathing pattern that gives away the underlying problem.

Eliciting peritonism can be performed by asking the child to cough, "blow out and suck in" their abdomen or gently percussing the abdomen. Rebound tenderness is unnecessary, uncomfortable and should not be performed in children; percussion tenderness is a suitable alternative. Asking the child to "jump up and down" is both fun for the child and is very reassuring if it is performed without causing pain; asking the child to jump on one leg only in turn can help to locate the pain to one side or other. A careful history of increased pain when 'the car went over bumps in the road' is indicative of peritonism and adds to the clinical findings and helps build the picture.

Per rectal examination is almost never indicated in a child with abdominal pain and should not be performed in the vast majority of cases.

Also, exposure and examination of the groin and the scrotum are an essential part of the initial examination. Listening to the chest is also useful in that basal pneumonia can present with abdominal pain and in case of peritonitis the child may not be able to perform breathing instructions of "breathe in and out" with expected degrees of freedom.

Differential Diagnosis

Table 19.1 presents the common causes of abdominal pain in children as per their age bracket. Whilst under 2 s can develop acute appendicitis, this is unusual and intussusception is more prevalent in this age group.

Appendicitis

The most common cause of abdominal pain that warrants prompt surgical management. The typical history is of central/periumbilical pain migrating to a position in the right iliac fossa over a period of 24–48 h. Often associated with vomiting, diarrhoea, anorexia and a low-grade temperature; however, there is not one symptom that clinches the diagnosis. A high-grade temperature more likely signifies a viral infection as a cause of the abdominal pain, unless the appendix has perforated in which case the

Table 19.1 Diagnoses to consider as per age group of child

0–2 s	2–12 s	12–16 s
Intussusception	Appendicitis	Appendicitis
Incarcerated hernia	Non-specific abdominal pain	Gastroenteritis
Colic	Gastroenteritis	Constipation
UTI	Constipation	Ovarian/testicular torsion
Notes: bilious vomiting warrants emergent surgical referral as may herald malrotation/volvulus of midgut	Others: tonsillitis, pneumonia, DKA, functional abdominal pain	Pelviureteric junction obstruction Inflammatory bowel disease Biliary disease Pancreatitis

abdominal findings should be unequivocal. Any child with acute pain in the right iliac fossa should have appendicitis considered. If the examination findings are of guarding in the right lower quadrant then prompt referral is recommended. However a retrocaecal appendix can lead to no overt peritoneal signs and so careful safety net advice is absolutely paramount with the child's carers knowing to re-attend if they are concerned that the symptoms are not settling or worsening over time.

Non-specific Abdominal Pain

This is the most common diagnosis eventually given to children who are referred to secondary care units with abdominal pain. Whilst often unsatisfactory to parents and clinicians, it is a useful umbrella term to describe the extremely common scenario of a child with sometimes quite distressing abdominal pain, usually few other symptoms, but who recovers without surgical intervention and simply requires analgesia to treat the pain whilst it occurs. It encompasses, and should be used in lieu of the frequently used diagnosis of "mesenteric adenitis", where the (often younger) child will complain of abdominal pain in association with an intercurrent illness (often upper respiratory tract) leading to the presumed swelling of the mesenteric lymph nodes and subsequent pain.

Intussusception

Age is the key differentiator here. Whilst over 2 s do get intussusception, it is far less common. The clinical triad of vomiting, pain and an abdominal mass is rarely seen together. However, the history is usually classical, with a prodromal illness leading to bouts of severe inconsolable crying, drawing up of the knees followed by periods of quiescence where the child is often pale and exhausted. The child's face usually goes pale during a crying episode, as opposed to red and flushed when secondary to screaming episodes from being unhappy for other reasons, eg constipation. Red currant jelly stools are a late sign, indicative of mucosal sloughing from the ischaemic bowel. Ultrasound is nearly 100% sensitive for this condition which can be treated with air enema reduction in many cases if caught early enough. Urgent referral is mandatory if this diagnosis is considered.

Others

Constipation is extremely common in children of all ages. The presumed colonic spasm that these children can experience can be very uncomfortable. Usually easily treated with a stool softener, occasionally it may require rectal medication to solve.

A good history usually makes this evident and palpable fecalomas in the left iliac fossa are not uncommon, sometimes this can be more extensive. Abdominal X-rays are usually not needed to confirm constipation, do not change management and expose to radiation and therefore are not recommended.

UTIs

It would be unusual for a UTI to present with abdominal pain alone, without lower urinary tract symptoms, however in younger children and toddlers in nappies it can be difficult to elicit frequency/urgency and dysuria cannot be clearly described by the child. A urine dipstick is an easy and useful test in all children with lower abdominal pain. Where UTI is suspected, in addition to a urine dipstick test the urine must be sent for culture and sensitivity tests.

Gastroenteritis

Diarrhoea and vomiting are often prominent features in acute appendicitis, especially in very young children and often there is diagnostic uncertainty. Gastroenteritis however should not lead to peritoneal signs on examination and often the GI upset is proportionally worse than the abdominal pain. Children are more vulnerable to fluid loss and if dehydration cannot be adequately countered through oral intake this should prompt referral to a paediatric unit.

Red Flags

A longer history with GI upset, weight loss and/or blood in the stools should lead you to consider the possibility of inflammatory bowel disease, especially in adolescent children.

Abdominal pain with a “mass” palpable is unusual. If it is indentable, with a history of constipation it is likely to be a faecaloma however outside of this scenario paediatric malignancy should be considered.

Bilious vomiting (dark green) is indicative of intestinal obstruction until proven otherwise and should be referred urgently for a surgical opinion.

Bruising of different ages on the abdomen, particularly in a withdrawn child with poor interaction with their carer raises the possibility of non-accidental injury and should prompt a thorough search for other injuries and referral to a child safeguarding team.

Beware the ‘viral illness’ with vomiting, high temperatures, abdominal pain and an irritable, immobile child under 5 years of age—acute appendicitis with early perforation and peritonitis is to be considered.

Abdominal pain with persistent diarrhoea and high temperatures and mild tenderness can be due to a deep pelvic appendicitis that has perforated and caused a pelvic abscess.

Pitfalls

These clinical situations are potentially mis-diagnosed:

Abdominal pain in the setting of under 5 s, children with autistic spectrum disorder or previously partially treated "UTIs" or "gastroenteritis" should warrant very cautious clinical evaluation. Appendicitis can masquerade in unusual ways but especially in these subgroups and referral, if any doubt, is advised.

Teenage girls with significant lower abdominal pain, particularly in the setting of vaginal bleeding should mandate a urinary pregnancy test. In all girls with severe lower abdominal pain ovarian torsion must be considered and emergent referral is recommended.

Always examine the groin of children with abdominal pain looking for incarcerated inguinal hernias and in boys always examine the testicles as referred abdominal pain from testicular torsion is a commonly missed diagnosis.

All physiological observations should be read in conjunction with the normal ranges for each age group. This will allow true physiological derangement to be identified early and "sick" children to be referred for prompt treatment in a paediatric or surgical unit if necessary.

Summary

Whilst the list of differential diagnoses is wide for each child presenting with abdominal pain, a thorough history and examination should help to narrow down the possibilities and identify the causes that require prompt secondary care referral.

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Chapter 20
Gastro-Oesophageal Reflux

David I. Campbell

Key Points
- Vomiting in young infants is common. The following should never be considered benign:
  - Bilious vomiting (dark green).
  - Haematemesis of more than a few streaks of blood.
  - Failure to thrive.
- A working knowledge of cow's milk allergy is important.

Introduction

Consider infants and young pre-school children as different from older children. The vomiting child is very obvious, but the consequences of reflux oesophagitis can be marked with a lesser grade of reflux (GORD without vomiting) (Table 20.1).

Investigations

Infants with symptoms suggestive of simple gastroesophageal reflux can be empirically treated with feed thickeners (alginate preparations such as Gaviscon®) or formula feeds changed with thickeners pre-added i.e. (Enfamil AR® Mead Johnson Nutrition). It is reasonable to empirically try an antacid (Ranitidine 2 mg/kg tds).

Children with eczema or strong family history of atopy, or with symptoms of fore gut disturbance (GORD) plus midgut disturbance (bloating, colic, iron defi-

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Table 20.1 Symptoms of Gastroesophageal Reflux and potential alternative diagnoses

(a) In the young child (<2 years)	
Young children	
Form of presentation	Differential diagnosis to consider
Profuse vomiting	3 months pyloric stenosis (dry FTT, inexorable history). UTI Raised intracranial pressure Metabolic diseases Part of cow's milk protein allergy
Failure to thrive	Due to calorie loss (i.e. vomiting, see above) Due to insufficient calorie intake (i.e. feed aversion (see below)). General medical clerking to identify other body systems involved. Always consider cystic fibrosis, coeliac in the weaned child and neglect.
Feed aversion	GORD may be co-existent with milk allergy Any cause of feed induced pain (peptic ulcer, small bowel stricture or ulcer, oesophagitis) May be an early presentation of autism
Feeding problems (gagging or choking)	Neurological problems such as CP/bulbar or pseudobulbar palsies Part of feed aversion (see above)
Haematemesis	Oesophageal varices Peptic ulcer Swallowed blood (pharyngitis or cracked nipples in breast fed babies).
Poor sleep	Raised intracranial pressure
Sandifer syndrome	Dystonia syndromes, epilepsy (rare situations)
Drooling	Causes of hypotonia
Colic and abdominal pain	UTI, constipation, milk allergy (often co-exists with GORD)
Chest infections	Causes of recurrent chest infection such as cystic fibrosis Primary immunodeficiency Congenital lung abnormality
(b) In the older child or adolescent	
Older child/adolescent	
Dyspepsia	Peptic ulcer disease, non-ulcer dyspepsia (gastritis), gallstones (cholecystitis), chronic pancreatitis
Waterbrash	Causes of upper GI dysmotility (coeliac, cholecystitis)
Vomiting/regurgitation	Migraine, oesophageal stricture General medical clerking to identify other body system disorders Post fundoplication (fundal wrap too tight)
Dysphagia	Oesophageal webs, peptic stricture (due to GORD usually), oesophageal tumours Eosinophilic oesophagitis
Nausea, halitosis	Any cause of vomiting
Laryngitis and dysphonia	Vocal cord palsy (consider raised intracranial pressure) or hypocalcaemia
Respiratory problems (dry cough)	Asthma Endobronchial infection Habit cough

ciency) or hind gut disease (diarrhoea) should be considered to have possible cow's milk protein intolerance. How these children should be treated will be dealt with elsewhere, but will include a dairy (or a dairy and soya free diet) together with the use of an extensively hydrolysed milk (Pepti® and Peptijuniour® both Danone Baby Nutrition or Nutramigen I or II® Mead Johnson Nutrition). More resistant cases (or on the basis of cost savings in some studies) may require an amino acid based formula (Neocate® SHS Nutrition) [1].

Children with troublesome Gastro-oesophageal reflux:symptoms or failure to thrive should have diagnosis confirmed, but also it should be defined what the cause of reflux is (hiatus hernia, dysmotility due to enteropathy, gastritis etc.). This should be done by endoscopy and ph study (catheter based as a combined impedance study or wireless Bravo Ph®).

Treatment

Empirical treatments can be continued if a positive response is seen.

We do not recommend more potent acid suppression (using a proton pump inhibitor) beyond 3 months duration without a confirmed diagnosis (see above). Treatment with any form of PPI beyond 3 months increases the risk of community acquired *Clostridium difficile* colitis, which can present acutely as a fulminant colitis [2].

PPI such as Omeprazole at 0.7 mg/kg od (max dose 20 mg day) in <2 year olds, or 10 mg od up to 20 kg body weight or 20 mg od for body weight above 20 kg. Higher doses should be used for shorter periods [3].

Indications For Paediatric Gastroenterology.

Severe disease with FTT or anaemia. Requirements for PPI. Confirm diagnosis. More complex food allergy than simple dairy intolerance.

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Chapter 21
Rectal Bleeding in Children

David I. Campbell

Key Points

- Consider the age of the child when considering the likely cause of a GI bleed.
- Looking for signs of upper GI bleeding is vital.
- Doctors should demonstrate they have considered haemodynamic stability as part of their assessment.
- Rapid gut transit time can cause upper GI bleeds to present as rectal bleeding.

General principles:

The following issues need to be considered in the history as they affect diagnosis and hence management:

1. Is this bloody diarrhoea?
2. Is this acute or chronic (acute on chronic)?
3. Are there any features to suggest upper GI bleeding (haematemesis and melaena)?

Sites of bleeding proximal to first loops of jejunum likely to have signs of upper GI bleeding, including melaena and haematemesis. Rapid transit would lead to bright red bleeding and can create the appearance of a lower GI bleed, even when the cause is from an upper GI source. This phenomenon is not uncommon in paediatrics. In addition there may be situations where there are multiple sites of GI bleeding, and this also is not uncommon in paediatrics (i.e. graft versus host disease,

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NSAID induced, coagulopathies or platelet related issues). Causes of upper GI bleeding vary from trivial (swallowed blood in breast fed infant, or nose bleeds to catastrophic haemorrhage from oesophageal varices). If in doubt consult with a paediatric gastroenterologist.

Consider the following differential diagnosis in

Older children: inflammatory bowel disease, infection, polyps, fissures.

Fissures: Usually secondary to constipation or idiopathic. If associated with induration, multiple or with skin tags, consider Crohn's disease. Also a feature of child sexual abuse. For unusual or multiple skin tags, or associated with anal or genital warts, consult with a paediatric colleague.

Neonates and Newborn

In newborns and young infants rapid transit can deliver unaltered blood to the terminal large bowel, obscuring the fact that the source of bleeding may be the upper GI tract.

Haemorrhagic disease of the newborn (HDN) is a vitamin K deficiency state, more common in breastfed infants and newborn. Bleeding can be from any site, but is often the GI tract. This condition is rare in babies given supplemental vitamin K at birth. HDN is a cause of haemorrhagic cerebrovascular accidents in young infants, hence an important diagnosis, irrespective of how much rectal bleeding has occurred.

Necrotising enterocolitis (NEC) is rare in term babies, but should be considered in any form of rectal bleeding in neonates. NEC is an idiopathic condition; most prevalent in the extremely premature when enteral feeding begins. Translocation of bacteria from gut lumen, vascular thrombosis leads to gut ischaemia and infarction with pneumatosis coli. When advanced the abdomen is discoloured, the baby shocked and peritonitic. Unless babies are fasted and treated with IV antibiotics, it progresses to intestinal perforation and peritonitis with a high mortality rate. Suspected cases should be referred to a paediatric surgeon.

Newborns with rectal bleeding may have rare diseases such as arterio-venous malformations affecting the GI tract, these babies usually have cutaneous haemangiomas as well. Most cases of rectal bleeding in new born should be investigated by a paediatric medical team.

Cow's milk protein allergy tends to produce colitic symptoms in a well child. Streaks of blood with mucous and loose stool. Children strain to pass a soft stool due to tenesmus and may have intercurrent reflux symptoms and colic. First line treatment is with an amino acid based formula if there has been rectal bleeding. Medical review is essential to confirm the diagnosis.

Intussusception (see below) is usually ileocolic and presents in an unwell child with colicky abdominal pain that turns pale during episodes of pain. The child may or may not be obstructed. Not every stool need be like redcurrant jelly (mucosal infarction), nor every stool bloody.

Infants and Young Toddlers

Defining whether the rectal bleeding is part of a bloody diarrhoeal illness is important.

Causes of bloody diarrhoea:

1. Infection (bacterial: *salmonella, shigella, campylobacter* and more notably *e. coli*). Usually early on there is a history of fever, pus or mucus are passed in the stool. Stools should be sent for microscopy and culture for three reasons, even if the treatment is conservative. Firstly, for public health reasons, secondly to identify specifically *e. coli 0157* which is a causative agent for haemolytic uraemic syndrome (HUS). Children with D+ HUS (i.e. HUS with diarrhoea, usually carry *e. coli 0157* and often need renal support or expert assessment by paediatric nephrologists. The final reason is that treatable causes such as *entamoeba histolytica*, are diagnosed on microscopy (and can be treated with oral metronidazole).
2. Dairy allergy (allergic enterocolitis). Usually these are well young infants that have mucousy, blood stained, loose frequent stools. The child usually is thriving and may be breast feeding. It is now known that microgram quantities of lactalbumin, lactoglobulin and casein proteins from cow's milk are found in human breast milk within a few hours of ingestion [1]. Any form of cow's milk infant formula will produce the same reaction in the susceptible infant. A 2 week dairy free diet using an extensively hydrolysed formula, or an amino acid based formula, is the gold standard test for dairy allergy [2].
3. Intussusception. This is a life threatening condition more common from 6 months of age, but can occur at any age (but the aetiology is different). Children have usually started weaning. The history is of episodes of crying (screaming) associated with pallor (rather than flushing which is normal). The child is unwell and the stools are classically described as redcurrant jelly. An abdominal mass on the right hand side is a classical finding, but smaller intussusceptions may not be palpable and an abdominal ultrasound should be employed to detect the apex of the intussusception. Children must be urgently referred to see a paediatric surgeon.
4. Perianal fissure. If the history is of a child straining to pass a large hard stool and intermittently there may be signs of bright red blood separate to the stool, then there should be a perianal fissure. Treat such children with laxatives.

Older Children

1. Inflammatory bowel disease. Either UC, Crohn's or indeterminate colitis will cause bloody diarrhoea. In all forms of IBD causing bloody diarrhoea the bleeding will be more prominent if the distal colon is affected. Associated symptoms of colitis/proctitis will be present: urgency, mucus, tenesmus, nocturnal diarrhoea, gastrocolic reflex (passage of stool after eating). Symptoms are chronic (or acute on chronic). Abdominal pain is a prominent feature.

2. Polyps. Painless rectal bleeding is the cardinal features on history. Usually a solitary juvenile polyp is the cause of painless rectal bleeding. Treatment is by therapeutic endoscopy and will require paediatric gastroenterology or surgical assessment. Rarely do forms of familial adenomatous polyposis cause bleeding.
3. Infection: as above
4. Perianal fissures. Treat with laxatives and GTN paste.

Conclusion

Age of child, presence of signs of upper GI bleeding and whether the bleeding is acute or chronic alter differential diagnosis and hence management.

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Chapter 22
Intestinal Obstruction

Jonathan Goring and Govind V. S. Murthi

Key Points

- Intestinal obstruction in children is not common but needs prompt recognition and management to avoid potentially significant complications.
- Abdominal pain, distension and vomiting are the commonest modes of presentation, with bilious vomiting particularly requiring urgent surgical assessment.
- Initial management should focus on fluid resuscitation and analgesia whilst investigations into the cause and definitive management can be arranged.

Introduction

Obstruction of the gastro-intestinal tract is one of the commonest reasons for a paediatric surgical opinion to be sought. All practitioners looking after children should be familiar with the common causes of intestinal obstruction and be aware of red flags that may herald a surgical emergency that warrants immediate referral and surgical management.

The causes of intestinal obstruction can be divided into:

- congenital causes - affecting mostly neonates in the first few days of life
- acquired causes - which are primarily diseases of infants and older children

The hallmarks of obstruction: abdominal distension, vomiting and abdominal pain will often be present. However, this may not always be the case, for example, interpretation of pain in a neonate can be difficult and proximal intestinal obstruc-

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tion will often present with a non-distended or even scaphoid abdomen. Therefore, diagnosing intestinal obstruction in children of different ages requires a knowledge of the possible causes combined with an appreciation of how each of these will likely present both clinically and radiologically.

Whatever the cause of obstruction, fluid loss both through vomiting and “third space” losses can mean these children can present to the clinician in a hypovolaemic state. Careful assessment of fluid status and initiation of fluid resuscitation is imperative whilst investigations and definitive management are organised.

Intestinal Obstruction in a Neonate

Assessing possible obstruction in a newborn baby focuses on congenital causes of intestinal obstruction, both mechanical and functional in nature. A standardised approach to these patients will help the practitioner consider the common causes and arrange appropriate management.

It can be useful to think anatomically (proximal to distal bowel) when thinking about causes of neonatal obstruction:

– Oesophageal atresia (with or without a trachea-oesophageal fistula)
  A very proximal cause of intestinal obstruction that presents within hours of birth with frothy oral secretions, inability to feed and dusky episodes on feeding.
– Duodenal Atresia
  Often picked up antenatally, can be confirmed radiologically with a “double bubble” sign seen on postnatal x-ray. Can present with bilious (post-ampullary obstruction) or rarely, with non-bilious (pre ampullary obstruction). If there is no antenatal history this presentation necessitates emergent exploration as it can mimic obstruction secondary to midgut volvulus. Otherwise cases can be dealt with “in hours.” One in three children with duodenal atresia will have trisomy 21 and an echocardiogram pre-operatively is recommended in all cases. Occasionally there is duodenal stenosis rather than atresia and these children may present a few weeks or months later with a history of recurrent vomiting and an enlarged stomach on x-ray.
– Malrotation and Volvulus
  Although this can present at any age, 70% present within the neonatal period. The malrotation of the bowel leads to a narrow mesenteric base which can then twist, culminating in bowel ischaemia and short gut or even death unless treated urgently. Bilious vomiting is the hallmark of this condition and necessitates an upper GI contrast to rule out malrotation. Abnormalities of rotation on the contrast study (Fig. 22.1) warrant prompt surgical exploration.
– Pyloric stenosis
  These infants present between 3 and 10 weeks of life with persistent, ‘projectile’, non-bilious vomiting. A background of vomiting from gastro-oesophageal reflux (much commoner) may be present but the persistent nature of the vomiting and also associated dehydration should raise suspicion and warrants a referral. Pyloric stenosis is easily diagnosed or excluded by means of an ultrasound scan (Fig. 22.2) and surgical correction is curative.

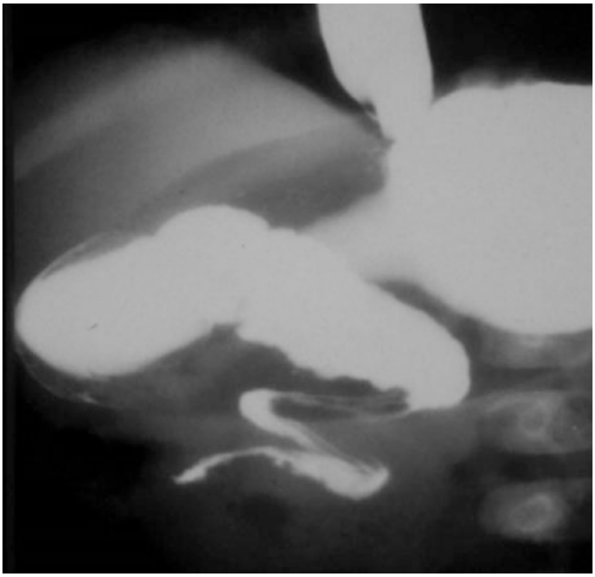

Fig. 22.1 Upper GI contrast study in a patient with malrotation showing spiral twist of the proximal jejunum indicating volvulus of the small bowel

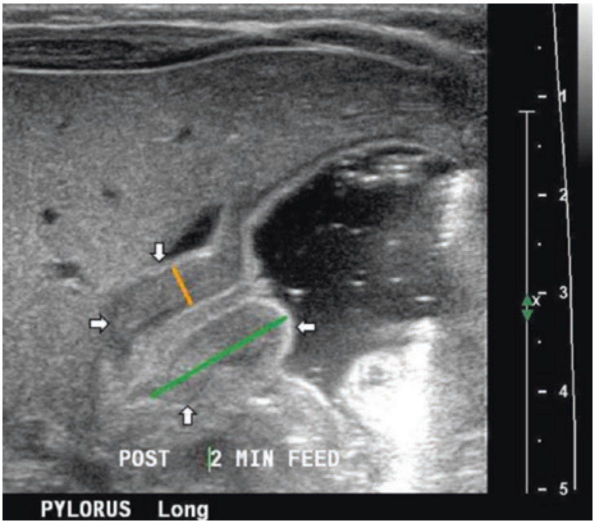

Fig. 22.2 Ultrasound picture of longitudinal section of pylorus in a patient with pyloric stenosis with arrows marking the length, width and pyloric wall thickness and also distended proximal stomach

- Ileal/Jejunal atresias
  Thought to be secondary to a late vascular event in utero affecting the mesentery to the bowel, they are often not picked up at early antenatal scans. This theory also explains why some of these children will pass meconium (present in the distal bowel before the obstruction occurred). Usually present with abdominal distension and vomiting in early neonatal life and large dilated loops of bowel on x-ray. Colonic atresia is rarer but presents similarly.

- Meconium Ileus
  The thickened putty like quality of meconium seen in children with cystic fibrosis (CF) will often obstruct the terminal ileum and colon. There may be an antenatal history of "echogenic bowel" or a family history of CF. The neonates may pass a whitish "plug" of meconium/mucous PR or nothing at all. In *simple* meconium ileus, a lower GI contrast study will often elucidate the pellets of meconium in a small, unused colon and can occasionally help to treat the obstruction by dislodging the meconium and allowing it to pass. If this fails, however, an operation is often needed. In *complex* meconium ileus (where bowel perforation, ischaemia or atresia is suspected) an operation is always required. Of note this is one of the few causes of a neonate being born with an already distended abdomen. CF genetics can be sent and if necessary, a sweat test arranged to confirm the diagnosis.
- Hirschsprung's disease
  Children with Hirschsprung's disease do not have ganglion cells in their distal bowel. The segment affected varies in length but is usually from the anus to the rectosigmoid area. As such the bowel cannot "relax" and is in a constant tonic state, leading to a functional obstruction and usually failure to pass meconium for the first 48hrs of life. Whilst a lower GI contrast study may help show the "transition" zone, where the normally ganglionated but dilated bowel meets the narrowed Hirschsprung's bowel, this is unreliable and a rectal biopsy is required to confirm the diagnosis in all cases. Initial management is with rectal washouts on a daily basis, but if this fails to relieve the obstruction the child may need a stoma pending further definitive surgery. Occasionally these children present very unwell with Hirschsprung's associated enterocolitis. In which case full resuscitation, including fluid, antibiotics and bowel washouts is vital as this can be a fatal disease if neglected.
- Anorectal malformations
- Affecting approximately 1:4000 live births, these children do not have a normal anus, instead the rectum ends blindly or as fistula somewhere in the perineum or urinary tract (boys only); in girls there is almost always an opening in the perineum – the anus may be in the vestibule or there may be only one opening in the perineum. There may be associated abnormalities as part of the VACTERL spectrum (Vertebral, Anorectal, Cardiac, Tracheo Esophageal, Renal, Limb) which should be actively looked for. Treatment for the majority of anorectal malformations requires a colostomy followed by definitive anorectoplasty later in infancy.

Questions to Ask

Several key pieces of information will help the clinician diagnose intestinal obstruction

1. Antenatal history.
   Most pregnancies in the UK will have had a 20-week anomaly scan. Abnormalities in the intestinal tract are often highlighted at this stage and these

children may be followed up with further imaging. “Echogenic” bowel is a very non-specific antenatal finding associated with CF but its positive predictive value is low. Polyhydramnios is often seen in proximal intestinal obstruction, however, is also seen in many other non-surgical conditions.

2. Has the child passed meconium?

   98% of term neonates will pass meconium within 48 hrs of birth. Neonates who present with vomiting and abdominal distension who have not opened their bowel by this time may have Hirschsprung’s disease, a distal atresia or meconium ileus. Occasionally this may be the presenting feature of an anorectal malformation

3. What colour was the vomit?

   Green vomit indicates intestinal obstruction until proven otherwise. In a neonate, especially, this can be a sign of malrotation leading to midgut volvulus. This is a time critical surgical emergency and requires immediate referral for a surgical opinion. Yellow vomit is common in infants and does not represent bile.

4. Is the neonate otherwise well? Any risk factors for sepsis?

   All neonates presenting with vomiting and abdominal distension should be treated for sepsis as a possible cause whilst other causes such as obstruction are ruled out. Risk factors for sepsis include prolonged rupture of membranes and maternal fever perinatally.

Assessing a Neonate with Possible Obstruction

A full clinical examination should be performed and possible sources of sepsis specifically looked for.

Specifically, the child’s abdominal examination should assess for distension, palpable loops or visible peristalsis which are often felt/seen underneath their thin abdominal wall. Inguinal hernias are a common cause of bowel obstruction and should be looked for in every case. It is vital to examine the perineum carefully. Meconium in the nappy at the newborn examination does not exclude an anorectal malformation (e.g./perineal fistula) and these abnormalities can be missed unless specifically looked for—one should ensure that any meconium smearing the perineum is wiped clean and the perineum is carefully inspected in a good light.

Investigations should include blood and urine cultures, inflammatory markers and a plain abdominal x-ray to assess the gas pattern. It is impossible to differentiate between small and large bowel and a guess regarding this aspect is best avoided. However, an estimate of “proximal” or “distal” obstruction can be gained from the amount of gas filled loops of bowel present. The abdominal x-ray can also help to rule out intestinal perforation. Most neonatal abdominal x-rays are performed with the child supine, assessment for free air can be difficult and occasionally requires a film to be performed with the neonate on its side as a lateral shoot-through.

If proximal obstruction is suspected, especially in the setting of bilious vomiting, an upper GI contrast study is warranted urgently to assess the rotation of the bowel.

Abnormal rotation on such a study combined with bilious vomiting warrants prompt surgical exploration to rule out a midgut volvulus. If a distal obstruction is more likely clinically and radiologically, a lower GI contrast is often more useful. Not only can it help with the diagnosis (e.g Hirschsprung's disease or meconium ileus) but it can also be therapeutic in the latter case, helping to washout the obstructing thick meconium and relieving the obstruction without recourse to surgery.

Acquired Causes of Intestinal Obstruction

These will present in children of all ages. Below is a summary of the commonest causes.

– Intussusception
  The commonest cause of intestinal obstruction between 4 and 36 months, these children will present with acute intermittent abdominal pain, often associated with vomiting, which may become bilious. The classical red-currant jelly stool of this condition is a late sign indicating likely bowel ischaemia. They are almost exclusively ileocolic in configuration and rarely the intussusceptum can pass all the way to the anus and be visible externally. There may be a palpable mass on examination. Whilst fluid resuscitation is the caregiver's primary goal, unless signs of peritonitis are present, air enema reduction has an approximately 70% success rate and should be attempted where facilities are available. If this fails, a laparotomy is warranted.
– Adhesional Obstruction (Fig. 22.3)
  Any child who has had an abdominal operation is at risk of forming peritoneal adhesions. These form between loops of bowel and other bowel loops, solid organs or the abdominal wall and can occasionally cause bowel to kink or twist leading to obstruction. Prior surgical history is vital in assessing children with intestinal obstruction and looking for scars on the abdomen may give a clue. If suspected, conservative treatment with an NG tube and IV fluids may be indicated initially but should be instituted under the care of a paediatric surgeon who will stay vigilant for signs of deterioration which may warrant surgical intervention.
– Appendicitis
  This common condition in children of all ages usually presents with an "acute abdomen", with signs of peritonism. Typically, the pain is peri-umbilical and then moves to the right iliac fossa and may be associated with vomiting. However, especially in younger children (less than five years of age) where the diagnosis can be difficult and is often therefore delayed, they may present with signs of small bowel obstruction secondary to an inflammatory phlegmon caused by a perforated appendix.
– Meckel's diverticulum
  These congenital remnants of the vitello intestinal duct may remain as a band from a Meckel's diverticulum to the umbilicus. The fixed band can lead to small

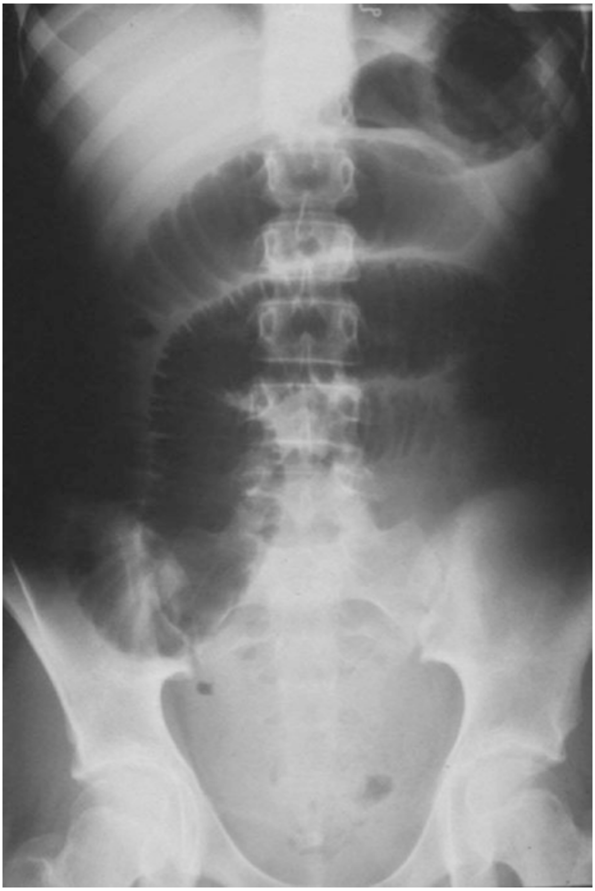

Fig. 22.3 Plain abdominal x-ray in a child with established small bowel obstruction showing dilated loops of small bowel as identified by the 'stacked coins' appearance of the mucosal folds

bowel twisting around it leading to acute small bowel obstruction, often in younger children.

- Constipation
  Constipation is common in children and can rarely present with features of intestinal obstruction (recurrent abdominal pain and vomiting). A good history and clinical examination where faecal masses can be palpable in the abdomen, especially the left iliac fossa, can help diagnose the cause. Rarely, pelvic masses with secondary constipation can also present with features of intestinal obstruction.

Is a per rectal examination necessary?

It can be safely said that at the initial assessment and examination by a general practitioner or paediatrician, a per rectal examination is best avoided—as it is often distressing to the child and family and uninformative.

Conclusions

Bowel obstruction in neonates, infants and children will usually present with abdominal pain, vomiting and abdominal distension; in neonates there is the additional feature of failure or delayed passage of meconium. A knowledge of the particular age-related causes can help the clinician to formulate an appropriate, logical management plan and understand the particular urgent "red flag" symptoms that necessitate immediate action to prevent catastrophic consequences. Bilious vomiting at any age, especially in a neonate and in infancy, warrants urgent referral to a paediatric surgical centre for further investigation and management. For the general physician or paediatrician who may be the first person to encounter the patient, it is important to remember that whilst the cause of obstruction may not be always evident at presentation, fluid resuscitation and pain control go a long way in ensuring a good outcome and should be commenced forthwith.

Suggested Reading

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Chapter 23
Bile-Stained Vomiting in the Neonate

Sean S. Marven and Prasad P. Godbole

Key Points
- Bile in vomit or gastric aspirate is green
- Any child with bilious vomiting/bile-stained aspirates should be referred to a paediatric surgeon urgently
- Malrotation with midgut volvulus is a life-threatening emergency

Introduction

It is normal for babies in the first few months of life to bring up small amounts of milk during or following feeds (possetting). As long as they are otherwise well and continue to grow then nothing further need be done. With earlier discharges following normal delivery (in some cases within 6 h of delivery), it is not unusual for newborns to present with bile stained vomiting and present to a specialist centre from home.

This chapter concerns bile-stained, i.e. green vomiting in the first month of life. Any neonate with bilious vomiting or bile-stained aspirates should be referred urgently to a paediatric surgeon to rule out an obstructive cause of the vomiting. Over one third of neonates admitted to a paediatric surgical unit with bilious vomiting may have a surgical cause [1]. Bile enters the gut in the second part of the duodenum and obstruc-

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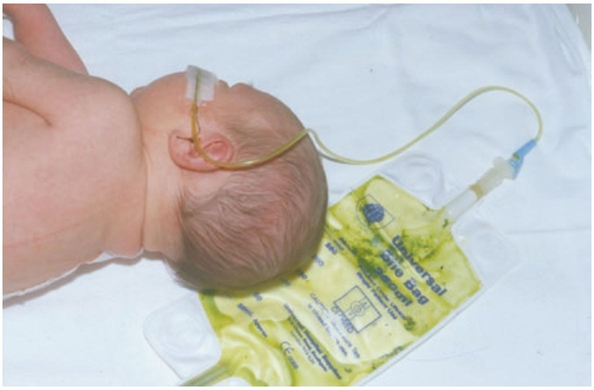

Fig. 23.1 Neonate with bilious aspirate

tion at any point below this level can result in bilious vomiting. It is possible that yellow vomit/aspirate may be seen early in intestinal obstruction but it would normally become green. Obstruction proximal to the second part of the duodenum, eg oesophageal atresia or pyloric atresia (rare) will therefore not cause bilious vomiting.

Amongst parents and healthcare professionals there is little agreement about the colour of bile in the newborn. When shown 4 shades of yellow and 4 shades of green as part of a questionnaire study, 56% of parents, 25% of GPs and 10% of midwives did not choose any of the shades of green as a possible match for bile [2]. It is therefore paramount when referred or seeing a neonate with reported vomiting that a clear history is taken from the parents or referring clinician about the actual colour of the vomit/aspirate. In the authors experience, parents often refer to normal gastric aspirate as 'bilious' (Fig. 23.1).

Assessment of the Neonate with Bilious Vomiting

Assessment of the neonate includes a detailed maternal, pregnancy and labour history with a search for clues that may give the cause for the vomiting. Points from the history include:

History	
Maternal • Medical conditions • Medications • Family history • Consanguinity Pregnancy • Antenatal scans – timing – bright or dilated bowel	Labour • Mode of delivery • Risk factors for sepsis • Condition at birth • meconium-stained liquor • green liquor (fetal vomiting) Early neonatal period • Vitamin K given? • Any feeds given/tolerated? • Delayed/failure to pass meconium • onset of vomiting/colour

The examination of the neonate includes an assessment of its general condition, may provide an estimate to the level of obstruction if present and a search for co-existing abnormalities:

Examination	
Respiratory condition Hydration/perfusion Jaundice Dysmorphic features Degree of abdominal distension	External appearance of perineum/anus Sacrum/spine normal? Hernia?

Initial investigations will be guided by your assessment of the child so far and the clinical setting you are in. The following should be considered but are by no means a prescriptive list nor exhaustive:

Investigations	
Bedside • urine dipstix • capillary blood gas • bilirubin Laboratory • full blood count (↓ platelets NEC/sepsis) • lactate (↑ NEC/ischaemia) • biochemistry (note renal function is maternal in first 24 h)	Radiological • supine plain abdominal x-ray • contrast studies (usually in tertiary paediatric centre • upper or lower GI contrast (ultrasound)

The purpose of the assessment is to determine the cause of the vomiting and guide initial management. The key features in neonatal bowel obstruction are:

- Bile-stained vomiting
- Abdominal distension
- Failure to pass or delayed passage of meconium

The level of obstruction determines the degree of abdominal distension. A neonate with a high obstruction may have no or a small degree of abdominal distension. They may also pass meconium initially and are likely to have onset of vomiting early. Conversely, a neonate with more distal obstruction will usually not pass meconium and may tolerate feeds initially. Over the first 24 h of life the abdomen will become progressively distended and they will then begin to vomit. At birth there is no air in the gastrointestinal tract but in a normal child it can take as little as 6 h for swallowed air to reach the rectum.

95% of term neonates pass meconium within the first 24 h and almost all by 48 h. Premature babies may have delayed passage of meconium. A neonate born in meconium-stained liquor has passed meconium but beware of green stained liquor. This may represent fetal vomiting of green bile into the amniotic fluid and therefore obstruction.

Neonates who may pass meconium but who may still have bowel obstruction include:

1. babies with Hirchsprung's may pass meconium, especially after rectal examination
2. some sticky meconium pellets may be passed in meconium ileus
3. onset of symptoms in malrotation with volvulus may be delayed for some time after birth [3]
4. a missed anorectal malformation with a fistula

Initial Management

At this stage the diagnosis may or may not be known but there are certain management steps that are common to all neonates with bilious vomiting:

Initial management	
Pass a nasogastric tube • 8 or 10 Fr in a term baby • aspirate and leave on free drainage (Septic screen ± lumbar puncture)	Commence iv fluids Commence iv antibiotics • as per local guidance

Causes of Neonatal Bilious Vomiting

The following is a (non-exhaustive) list of causes of bilious vomiting in the neonate. If a malrotation with volvulus is missed then the consequences, i.e. dead midgut, are disastrous and it is for this reason that any child with bilious vomiting should be referred urgently to a paediatric surgeon.

Causes	
Medical • necrotizing enterocolitis • sepsis – urinary tract infection – meningitis – group B streptococcus • raised intracranial pressure • hypothyroidism • prematurity • idiopathic bilious vomiting	High obstruction • malrotation ± volvulus • duodenal atresia/stenosis • proximal jejunal atresia Low obstruction • Hirschsprung's disease • ileal/colonic atresia • anorectal malformation • meconium ileus • meconium plug syndrome • small left colon syndrome • hernia

Malrotation With/Without Midgut Volvulus

Malrotation is failure of the gut to complete anti-clockwise rotation resulting in the entire small bowel lying attached by a narrow stalk rather than a broad based mesentery to the posterior abdominal wall. It is therefore prone to twist around on this stalk which compromises the blood supply (superior mesenteric vessels) of most of the small bowel—a midgut volvulus. Up to 80% of cases of malrotation present in the first month of life and most of these in the first week [4]. Therefore a previously apparently normal neonate (or child of any age) can present with a midgut volvulus. This is one of the reasons for urgency of referral in bilious vomiting. If a midgut volvulus is missed then the entire midgut can become unviable. At best this leads to short gut and lifelong parenteral nutrition, at worst, death. Plain abdominal x-ray may show dilated stomach and proximal duodenum but may also be entirely normal in malrotation. The diagnostic modality of choice is an upper GI contrast study. This demonstrates the duodenal-jejunal flexure lying to the right of the spine and inferior to the duodenal bulb. Treatment involves urgent laparotomy with resection of any dead gut and Ladd's procedure (Figs. 23.2 and 23.3).

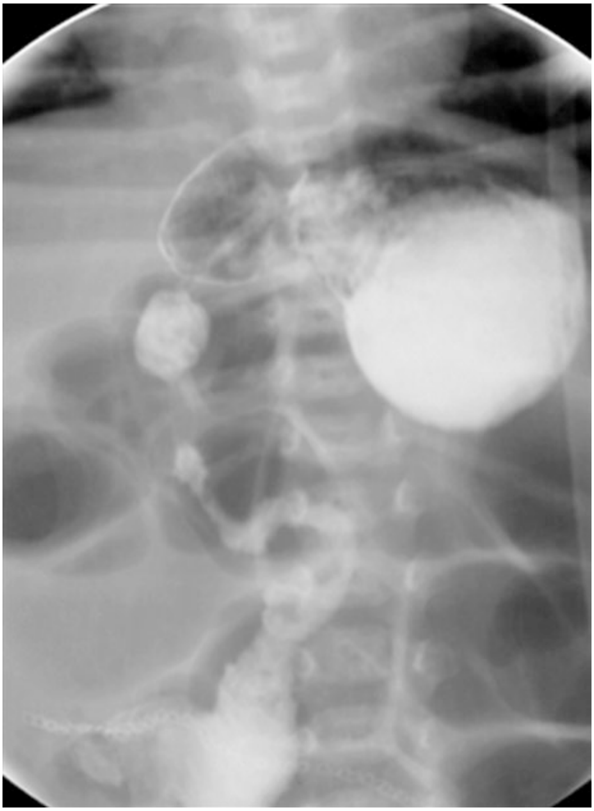

Fig. 23.2 Upper gastrointestinal contrast study demonstrating malrotation. The duodenal-jejunal flexure lies to the right of the spine

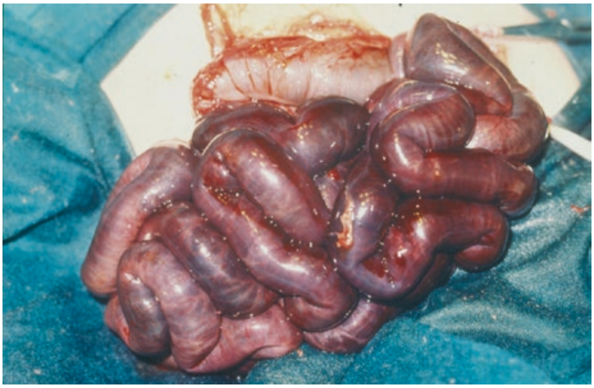

Fig. 23.3 Dead gut found at operation for malrotation with midgut volvulus

Necrotising Enterocolitis

This is almost exclusively a disease of preterm infants (although it can occur at term) and is therefore uncommon outside of the neonatal unit. Necrotising enterocolitis (NEC) consists of bowel wall inflammation and ulceration which may proceed to perforation. Neonates present generally unwell with apnoeas and bradycardias, green vomiting and temperature instability. The abdomen may be distended and shiny or erythematous and there may be rectal bleeding. Abdominal radiograph features include distended bowel loops, intramural gas (*pneumatosis intestinalis*), portal vein gas and pneumoperitoneum. Management is mostly undertaken by the neonatology team and consists of stopping enteral feeds, a nasogastric tube on free drainage and antibiotics. Complicated NEC may require surgical intervention including perforation and stricture formation. Mortality for NEC with perforation remains between 30% and 50% [5].

Duodenal Atresia

Traditionally it is reported that a third of infants born with duodenal atresia have Down syndrome [6]. With a decrease in the number of Down children being born, this figure may be as low as 1 in 10. Conversely, 1 in 20 Down infants are born with duodenal atresia [7]. Approximately half of cases are diagnosed antenatally with the appearance of a fluid-filled double-bubble on ultrasound. Over half have associated congenital abnormalities including Down's syndrome, cardiac anomalies, VACTERL, malrotation and other bowel atresias [8]. Vomiting may or may not be green depending on the site and configuration of the abnormality. The typical appearance on plain abdominal film is a double-bubble which represents a dilated,

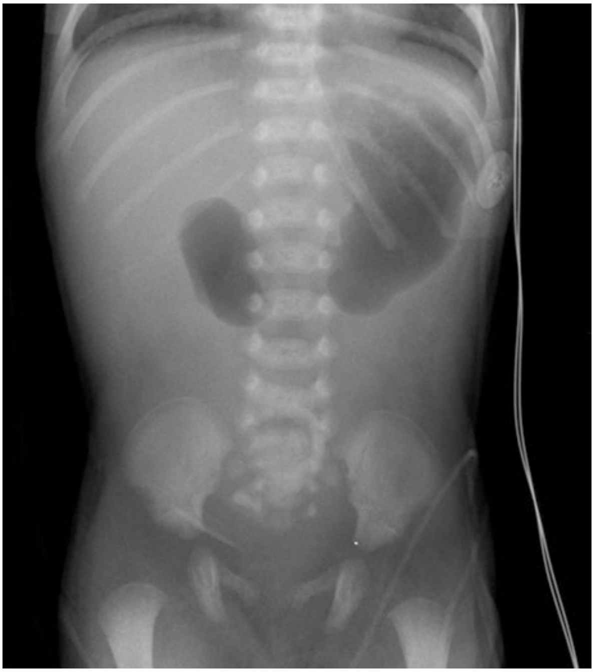

Fig. 23.4 Plain abdominal film demonstrating typical double-bubble appearance in duodenal atresia with no distal gas

air-filled stomach and duodenum. If there is stenosis causing the obstruction, there may be distal air also present on the plain film. However, in this case it is important to consider that malrotation may be the diagnosis. Surgical treatment involves creating an anastomosis between the proximal and distal duodenum without disturbing the atresic or stenotic segment as the common bile duct commonly enters the bowel here (duodenoduodenostomy) (Fig. 23.4).

Small Bowel Atresia

Atresias may occur more distally in the jejunum, ileum or rarely in the colon. There may be a single atresia, a large atresic segment like an apple-peel or multiple atresias. The level of the most proximal atresia determines the degree of abdominal distension, the timing of onset of vomiting, and the number of distended bowel loops that will be evident on plain abdominal film. Associated abnormalities include gastroschisis, cystic fibrosis and cardiac anomalies as well as atresias in other parts of the GI tract. Surgical management involves resection of the atresic segment and either primary anastomosis or temporary stoma formation. Extensive atresia(s) may leave a child with short gut syndrome (Fig. 23.5).

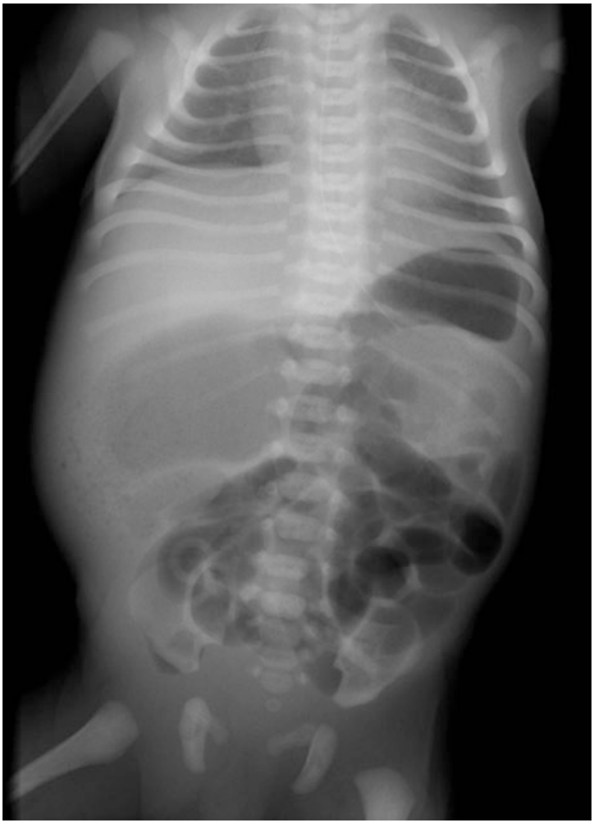

Fig. 23.5 Plain abdominal film of distal atresia (colonic atresia)

Meconium Ileus

Meconium ileus is the presenting feature in up to 1 in 5 children with cystic fibrosis [9]. However, 1 in 5 of neonates presenting with meconium ileus will not have cystic fibrosis [10]. This condition occurs when abnormally thick meconium impacts in and obstructs the distal ileum. Abdominal distension is a key feature and plain abdominal film may show a soap bubble appearance of the meconium. Diagnosis and treatment is with (high osmolar) water-soluble contrast medium. Should this fail then surgical intervention is required. Interestingly, the long term pulmonary outcome of cystic fibrosis children does not differ between those who present with meconium ileus and those who do not [11].

Hirschsprung's Disease

Agangliosis of a continuous segment of the large bowel extending for a variable distance proximally from the rectum leads to a functional bowel obstruction. The aganglionic bowel maintains a tonic contraction with dilated normal bowel proximal to this. Delayed passage of meconium or constipation in the first 4 weeks of life should raise the suspicion of Hirschsprung's disease [12]. Contrast enema may be suggestive but diagnosis is confirmed by rectal suction biopsy demonstrating an absence of ganglion cells and presence of acetyl cholinesterase (AChE)-positive hypertrophic nerve fibres. Initial management involves rectal washouts (proximal to the contracted bowel) to decompress the bowel or stoma formation until definitive procedure(s).

Anorectal Malformations

Babies with early discharge after birth may go home with an unrecognised anorectal malformation and it is vital that a proper inspection of the perineum and anus occurs before discharge. The passage of meconium does not exclude abnormal anatomy as there may be a fistulous tract with the rectum communicating with the urinary tract in boys, the vestibule or vagina (as a cloaca) in girls or the perineum in either. The finding of an anorectal malformation should trigger a search for associated abnormalities, particularly belonging to the VACTERL association (V-vertebral, A-anorectal, C-cardiac, TE-tracheo-oesophageal, R-renal, L-limb). After a thorough pre-operative work-up, initlal surgical management involves either a procedure to restore correct anatomy or a defunctioning colostomy.

Conclusion

Bile-stained vomiting in the neonate may herald intestinal obstruction. Delay in referral or in diagnosis may result in perforation, ischaemia or death. Happily, all the anomalies may be corrected with little or no long term effects. The dictum 'Bile stained vomiting in the neonate is surgical until proven otherwise' should be adopted to ensure timely management.

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Chapter 24
Unique Considerations in the Infant and Neonate: Pyloric Stenosis

Charlotte Holbrook and Joseph Curry

Key Points
- Pyloric stenosis presents as non bilious vomiting which is described as projectile.
- Associated biochemical anomalies includes a metabolic alkalosis and electrolyte disturbances.
- Diagnosis is by palpating the typical 'olive' in the epigastrium or by ultrasound.
- Surgery either open or laparoscopic is curative.

Introduction

Pyloric stenosis is a common pediatric surgical condition with an incidence of 2–4 per 1000 live births [1]. It affects males five times more than females [2]. The etiology of this condition is unclear. It is postulated that prolonged contraction of the pyloric muscle causes hypertrophy of the muscle fibres and development of a pyloric mass. One theory is that this occurs due to failure of relaxation of the pyloric muscle secondary to abnormal innervation, involving the nitric oxide pathway [3]. An alternative hypothesis is that excessive pyloric sphincter contraction occurs due to hyperacidity within the duodenum. High levels of gastrin have been documented after birth in some patients with pyloric stenosis [4]. A Danish nationwide cohort study demonstrated a 30-fold increased risk of developing HPS with use of prokinetic macrolide antibiotics in early infancy [5]. There is a definite genetic component to the condition. 1 in 20 of male offspring and 1 in 50 of female offspring from a male parent with pyloric stenosis will be affected [6].

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Presentation

The diagnosis of pyloric stenosis can often be elicited from the history. It typically presents as non-bilious projectile vomiting after feeding in infants between 2 and 12 weeks of age, with peak incidence at 5 weeks [4]. In the initial stages of the condition the babies are well and hungry to feed. As the condition progresses they become dehydrated and demonstrate weight loss. Common conditions mimicking pyloric stenosis are gastro esophageal reflux (GOR) and overfeeding. The vomiting may be projectile in gastroesophageal reflux, but the dehydration and weight loss noted in the later stage of pyloric stenosis does not occur. A careful feeding history is helpful to identify if the vomiting is from over feeding or feeding too concentrated a formula. The other important point to elicit is a family history of pyloric stenosis on either the maternal or paternal side.

Examination of a child with suspected pyloric stenosis should begin with an assessment of their hydration status. Increased capillary refill time, sunken fontanelle, loss of skin turgor and reduced alertness are all signs of dehydration occurring secondary to persistent vomiting of feeds. The classical abdominal signs in a child with pyloric stenosis are the presence of reverse gastric peristalsis on inspection of the abdomen and palpation of an olive shaped mass in the right hypochondrium. Palpation of the pyloric mass requires time and patience. The baby needs to completely relax their abdominal musculature. This can be facilitated by giving the baby a feed and flexing the hips during the examination. Palpation from the left hand side is also helpful as this helps to avoid the caudate lobe of the liver, which can sometimes be mistaken for the pyloric mass.

If pyloric stenosis is suspected from the history, blood should be taken for blood gas and biochemical analysis. Pyloric stenosis results in a hypochloraemic hypokalemic metabolic alkalosis. Alkalosis and hypochloraemia result from vomiting of gastric fluid rich in hydrogen and chloride ions. Dehydration from fluid loss leads to a release of aldosterone to maintain intravascular volume by reabsorption of sodium from renal tubules in exchange for potassium ions (leading to hypokalaemia). High bicarbonate levels are present due to reabsorption along with sodium ions, exacerbating the alkalosis. As the hypokalaemia worsens, potassium may be retained in exchange for hydrogen ions resulting in paradoxical aciduria. In the initial stages of the condition the biochemical abnormalities may not be present as the body compensates by producing more alkaline urine. Unconjugated hyperbilirubinaemia is present in 2% of patients due to a deficiency of hepatic glucuronyl transferase which resolves post operatively [7].

While history and clinical examination are often sufficient to confirm the diagnosis of pyloric stenosis, a pyloric ultrasound has become part of the work up for a vomiting baby in many pediatric surgical units. Pyloric ultrasound performed by an experienced observer has a sensitivity and specificity approaching 100% [8]. There are studies that have shown that fewer infants present with the classic palpable 'olive' or severe electrolyte disturbance [9, 10], which may be due to the more widespread use of ultrasound. The signs of pyloric stenosis on an ultrasound are a pyloric muscle thickness >3 mm, an elongated pyloric channel length of >15 mm and failure of relaxation of the pyloric canal (Fig. 24.1). There is a cor-

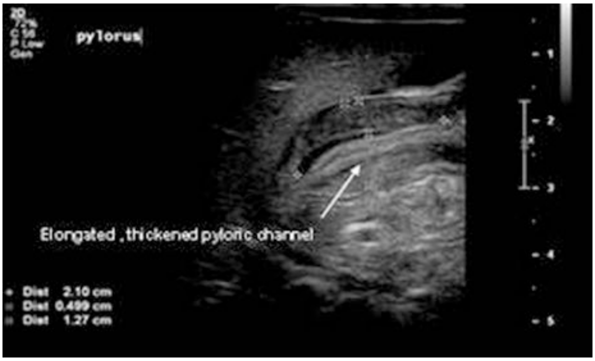

Fig. 24.1 Appearance of hypertrophic pylorus on ultrasound scan

relation between patient weight and age; up to 5% with pyloric stenosis will have a pyloric thickness of <3 mm [11]. UGI contrast has now been superseded by ultrasound as the radiological investigation of choice where the diagnosis of pyloric stenosis is suspected.

Management

The mainstay of treatment in pyloric stenosis is correction of electrolyte abnormalities prior to definitive surgical treatment. Intravenous fluids (0.45% or 0.9% sodium chloride and 5% dextrose) should run at 120–150 ml/kg/day with supplemental potassium added to correct the hypokalemia (10–20 mmol/L). This can be reduced to 100ml/kg/day when serum bicarbonate is <25 mmol/L [12]. An NG tube should be passed, aspirated regularly and losses replaced with intravenous 0.9% normal saline and supplemental potassium. The baby should be fasted until surgery. Venous blood gases and electrolytes should be measured approximately every 8 h until they have normalized. Failure to correct electrolyte abnormalities prior to surgery can lead to myocardial dysfunction following anesthetic induction and respiratory depression post operatively [13].

Surgical techniques in the management of pyloric stenosis have evolved since the modern day operation was first described by Ramstedt in 1912. While the main aim of the surgery, i.e., extra mucosal splitting of the pyloric muscle, has not changed, the approach has modified. The classic approach is a right upper quadrant incision to gain access to the pylorus. In the 1980s a circumumbilical incision was described to deliver the pylorus [14]. This technique grew in popularity due to the excellent cosmetic result post operatively. The first laparoscopic pyloromyotomy (Fig. 24.2) was reported in 1991 and has now been adopted by many centres as the technique of choice [15]. A large multicentre randomized control trial demonstrated a shorter hospital stay and quicker time to full enteral feeds using the laparoscopic approach over the circumumbilical approach [16]. A recent large retrospective comparison of the two approaches also demonstrated a statistically significant reduction in composite morbidity with the laparoscopic approach [17].

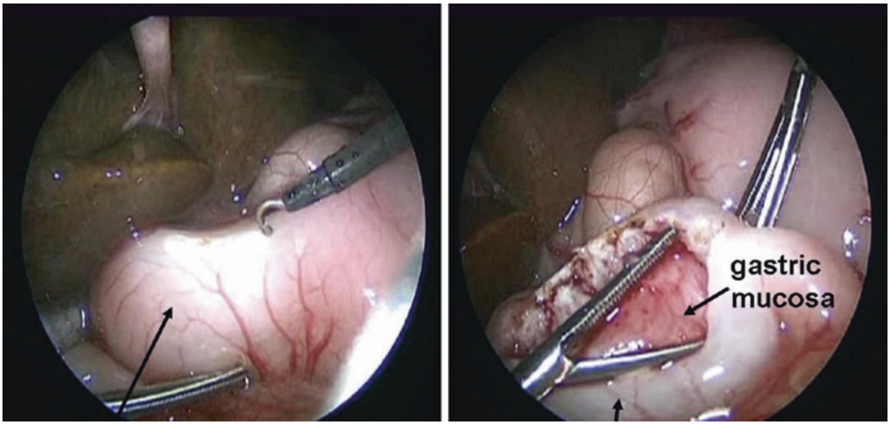

Fig. 24.2 Laparoscopic appearance of pyloric mass and pyloromyotomy

Feeding can begin approximately 6 hours post-surgery. Feeds can be built up incrementally or given 'ad lib': there is no evidence that either approach is superior [18]. Vomiting is common in the first 24 h post-surgery and parents should be reassured. Children are discharged once full feeds are tolerated which is usually on the second post-operative day.

Overall morbidity rates from pyloromyotomy vary between 4.6% and 12% [12] Complications include: intra operative mucosal perforation, wound infection, wound dehiscence and inadequate splitting of the pyloric muscle resulting in prolonged vomiting and recurrence of pyloric stenosis [19]. Mortality from pyloric stenosis is rare and results from an unrecognized mucosal perforation and subsequent leak of gastric contents.

Medical therapy using IV and oral atropine to treat pyloric stenosis has not gained widespread acceptance due to the longer length of hospital stay required and up to a 25% failure rate associated with the treatment [20]. Long term follow up of babies with pyloric stenosis demonstrate no significant side effects or symptoms from this condition.

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Chapter 25

Haemangioma, Lymphangioma and Vascular Malformations

Ross Fisher and Sarah Blundell

Key Points

- Haemangiomas usually appear in infancy, most are benign and will resolve, only requiring intervention if bleeding or large in size and compromising airway or vision.
- Kasabach-Merritt phenomena is a rare life threatening problem that occurs with only 2 types of rare infantile haemangioma which can lead to disseminated intravascular coagulation.
- Lymphangiomas usually affect the head and neck but can occur anywhere. Treatment is required if the lesion is obstructive (airway or intraabdominal) or causes cosmetic distress to patient or parents.
- Venous and arterial malformations with require multidisciplinary team management if intervention is indicated.
- It is important to consider associated syndromes and extent of malformation in children presenting with vascular anomalies.

Introduction

Anomalies of capillaries, arteries, veins and the lymphatic vessels are relatively common with most presenting in childhood. Some will resolve with time and others require intervention. Lymphangiomas and vascular malformations arise due to disordered formation of those vessels, haemangiomas are vascular tumours which can be sub classified into benign and malignant (not dealt with in this chapter). The

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majority of vascular anomalies found in children require reassurance only, if treatment is indicated referral to a tertiary level paediatric unit with surgical, medical and radiology expertise is usually necessary.

Common Conditions

Haemangioma

Infantile Haemangioma: benign hamartomas that present in the first few months of life with a period of rapid growth before involution usually within 5 years (Fig. 25.1). May range in size from small Strawberry naevi to giant infantile haemangioma. Rarely may present at birth as Congenital haemangioma.

Strawberry naevi: superficial, raised lesions within the skin that undergo involution in patches, hence “strawberry” nomenclature.

Pyogenic granuloma: A misnomer! Neither a granuloma nor pus producing but friable capillary haemangiomas at sites of trauma (or umbilicus) with a tendency to bleed on minor trauma.

Port wine stain is a capillary malformation, present from birth as a flat red lesion. They can be present in any location but are most common in the head and neck areas. They do not involute. May be representative of a deeper vascular malformation involving other structures such as the trigeminal nerve or spine.

Kassabach-Merritt phenomenon is associated with with 2 specific types of rare malignant infantile haemangioma (Tufted angioma or kaposiform haemangioendothelioma) [1] and results in thrombocytopaenia, hypoalbuminaemia and anaemia. The sequestration of clotting factors and platelets within the lesion this can lead to disseminated intravascular coagulation.

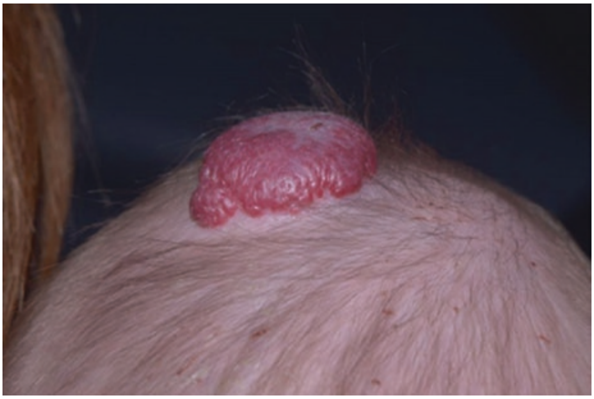

Fig. 25.1 Infantile haemangioma (a large strawberry naevus) on the scalp of a 6 month old child

Lymphangioma

A further incorrect term representing disordered formation of lymphatic vessels that may be sporadic or associated with a genetic condition. Most commonly affect the head and neck, they can also present as tumours of the limbs, mediastinum or abdomen (intra and retro-peritoneal). They are classified by size of the lymphatic cysts that are present, as macrocystic, microcystic and combined [2]. Ultrasound scan (USS) is the initial imaging of choice and use of Doppler can differentiate lymphangiomas from vascular malformations.

Vascular Malformations

Venous malformations can vary in size and location (affecting intraabdominal organs as well as skin) and although present from birth can become more evident with time. When examining the lesion they will increase in size with dependency and the Valsalva manoeuvre. These are known as “low flow” lesions. They are commonly symptomatic, causing problems by pressure or mass effect, haemorrhage or coagulation [3].

Arterio-venous malformations are present from birth and can occur at any site. They commonly effect the head and neck, both extra and intracranial. They represent an abnormal shunt between the venous and arterial systems and are therefore under pressure and are known as “high flow” lesions. Despite being present at birth, they may only be noticed as childhood progresses and the lesion grows. On examining these lesions there may be a palpable pulse or thrill as well as warmth to touch. In later stages they can cause local ulceration, tissue necrosis and heart failure.

Combined lesions: There are many syndromes which involve combined vascular lesions, such as Klipple-Trenauny-Weber syndrome.

USS with Doppler is usually the initial imaging of choice, progressing to MRI or angiography if required.

Treatment

Haemangiomas

Infantile haemangioma (eg Strawberry naevi): Watch and wait. Involution will occur, showing clearing and fading from the centre of the lesion outwards (Fig. 25.2). Larger lesions may be treated with propranolol (no one knows the exact mechanism of action of this treatment but it is thought to be due to a reduction in growth factors with an increase in haemangioma endothelial cell death) [4].

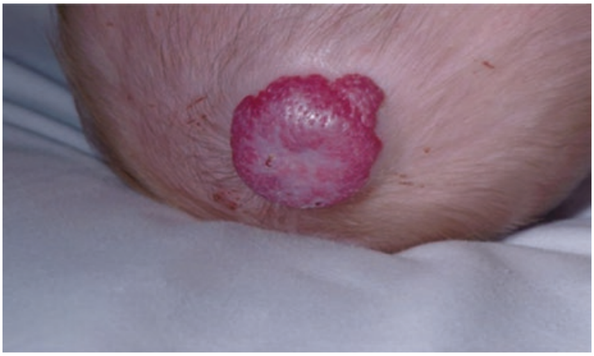

Fig. 25.2 Involuting infantile haemangioma showing central clearing

Pyogenic granuloma: silver nitrate, surgical excision.

Port-wine stains: depending on site and effect on the child: laser removal, coupled with investigations for associated syndromes (eg intracranial vascular malformation associated with port wine stain in the distribution of the trigeminal nerve in Sturge-Weber syndrome).

Salmon patches-remain present but become less noticeable with time-reassurance is all that is required.

Lymphatic malformation: Treatment for these depends on the site, the size, the effect on the child and the classification. For macrocystic lymphangiomata a sclerosant such as Doxorubicin, bleomycin or OK-432 can be used [5]. OK-432 is inactive Group A Streptococcus Pyogenes that is unable to replicate but does have antineoplastic capabilities (it was originally developed and used in the treatment of cancer). It creates an inflammatory reaction followed by fibrosis [6]. This can be followed by surgical excision or surgical excision alone can be attempted for localised lesions.

Venous malformations: A multidisciplinary approach is appropriate for venous malformations. A combination of medical therapies (compression garments, anti-inflammatories and anti-coagulants), with schlerotherapy (often US guided and with the use of tourniquets to limit the systemic uptake of the sclerosant). The sclerosants used differ from those used in lymphatic malformations and can include sodium tetradecyl sulfate or bleomycin [7]. Surgical excision of limited lesions or laser therapy for superficial lesions can be used [7].

Arterio-venous malformations: The treatment depends on the type of AVM present; either focal (one feeding vessel) or multicentric (many feeding arteries). The natural history of AVMs is for them to progress through stages in time, causing increasing amounts of tissue destruction to its surroundings. This mandates that all AVMs be treated. Embolisation of the nidus or draining veins of the lesion is the mainstay of treatment. This can utilise coils or polymers to create a blockage. Surgical excision is possible for focal lesions but often compromised by poor wound healing [7].

Indications for Referral

This depends on the lesion, the site and the symptoms experienced by the child and wishes of the family. And obviously depends on the experience and confidence of the clinician they present to.

Lesions that compromise the nose, mouth and neck that may have an effect on breathing need urgent referral; this may mean antenatal counselling and planned delivery in a tertiary centre for immediate surgical and airway management.

Lesions that effect vision or mobility would also mandate urgent referral.

Lymphatic malformations affecting the abdomen may be referred urgently if they present as an abdominal mass or compromise of mesenteric blood flow.

Bleeding, ulceration, rapid growth, failure to involute or emotional distress for the children would all be indications for referral for assessment and further management.

Referral to the appropriate tertiary service is also important.

Haemangiomas requiring propranolol treatment are often treated by plastic surgery, dermatology, or paediatricians. Lesions requiring laser removal are often managed by plastic surgeons in paediatric centres. Lesions such as arterial, venous or lymphatic malformations tend to be managed of initially seen by the surgical team which operates on that area of the body eg head and neck: ENT. Thorax/abdomen: paediatric surgery. However there is significant cross over in the services provided at different tertiary paediatric centres in the UK.

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Chapter 26
Rectal Prolapse and Pilonidal Sinus

Richard Lindley

Key Points
- Rectal prolapse is common in children and usually responds to conservative management with laxatives.
- Look for rare causes of rectal prolapse.
- Refer refractory cases for further treatment.
- Pilonidal disease in children is relatively rare but difficult to treat.
- Refer acute pilonidal abscess urgently for surgical treatment.
- Suspected pilonidal disease in pre-pubertal children is very rare and more likely to be a different pathology.

Rectal Prolapse

Introduction

Rectal prolapse in children is relatively common, benign and self limiting. It rarely discomforts the child but usually produces considerable anxiety in the carers. Managing this anxiety is often the most challenging aspect, as the majority of cases could be successfully dealt with in primary care. There are rare but important underlying causes that should be looked for and refractory cases will need onward referral. The highest incidence is said to be in infants and children below the age of 4, and in children girls are more commonly affected than in adult practice, although there is still a male preponderance.

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Pathophysiology

Typically, rectal prolapses are classified as partial (mucosal only) or full (mucosa and rectal wall); with the degree of full prolapse being further subdivided. Most paediatric rectal prolapses are full thickness but in practice the distinction in primary care will not affect the initial management or referral.

Prolapse in infants is most likely to be due to anatomical considerations, such as the shape of the pelvis and the attachments of the rectum and sigmoid colon. It will usually resolve spontaneously.

The vast majority of rectal prolapses in older children will be due to underlying constipation and/or disordered defaecation and respond to treatment of these problems. The challenge is to identify the small number that are caused by more serious underlying pathologies, which include:

- Cystic fibrosis
- Dysfunctional urinary voiding
- Neurological conditions
- Malnutrition
- Connective tissue disorders
- Hirschsprung's disease

An internal rectal prolapse which does not protrude beyond the anus (internal intussusception of the rectum) usually presents with rectal bleeding and problems with bowel function. These are difficult to diagnose in primary care and would usually be referred due to persistent bleeding.

Diagnosis

The presentation of rectal prolapse is usually dramatic and distressing to the child's carers. Occasionally the prolapse will still be present but usually it will have resolved spontaneously by the time a clinician is consulted.

The differential diagnosis is usually limited, as prolapsed haemorrhoids do not occur in the paediatric population. It is important to distinguish a prolapsed rectal polyp, which is a fleshy lump on a stalk, from the complete ring of a rectal prolapse. The ubiquity of smartphones with cameras can help with this if the carers are encouraged to take a picture of the problem when it occurs.

Once a prolapse is diagnosed, the rest of the history and examination should be directed at establishing the underlying cause.

Look for:

- Constipation
- Straining at stool
- Recurrent respiratory problems
- Failure to thrive
- Urinary problems (excessive straining or incontinence due to neurological problems)
- Lower limb neurology
- Failure to pass meconium within 48 h of birth

No further investigations should be necessary in primary care.

Primary Care Management

If constipation and or straining at stool are diagnosed, then simple laxative treatment is usually successful in treating rectal prolapse in children. An adequate stool softener, such as polyethylene glycol 1-2 sachets per day and titrated to response to allow easy passage of a soft stool, will suffice. Treatment for at least 2–3 months is usually required.

The carers may also need education in how to gently manually reduce the prolapse if it does not resolve spontaneously; this is done with gloves, lubricant and steady backwards pressure. If the prolapse has become oedematous then copious amounts of table sugar can be applied for 10 minutes to reduce the swelling.

When to Refer

Refer to a paediatric surgeon with an interest in colorectal problems when an underlying cause other than constipation or straining is suspected, or when simple laxative therapy has failed.

In the era of neonatal screening for cystic fibrosis it is unusual for a rectal prolapse to be the presenting feature, but in a child with other features, such as recurrent respiratory problems outside of the norm for childhood, or failure to thrive, it should be considered. Likewise, children with a suspicion of lower limb neurology should be referred for further investigation.

Surgical treatment options include injection sclerotherapy, per rectal resection and suturing techniques and laparoscopic hitching of the rectum with or without an implanted mesh.

Pilonidal Sinus Disease

Introduction

Pilonidal disease is relatively rare in children, with a reported incidence of about 1–2 per 1000, and it is almost exclusively a disease of post-pubertal children. A diagnosis of pilonidal disease in a pre-pubertal child is likely to be incorrect: other pathologies such as a natal cleft sinus or dermal sinus are more likely in this age group and tertiary referral should be considered.

Pathophysiology

Classically, pilonidal disease is attributed to broken and ingrowing hairs in the natal cleft and can therefore be expected to be more common in hairy adolescent males, but in fact hairs shed from the head in girls have also been implicated as they fall down the back and get trapped in underwear, meaning that the male:female sex ratio in children is more equal than in the adult population.

Once the hair begins to ingrow, a pilonidal sinus will form. These are epithelialized tracts that extended ever deeper. They can be asymptomatic, but if they become infected pain and discharge will occur. As the disease progresses subcutaneous infection can result in abscess formation. Eventually large chronically infected subcutaneous cavities with “nests” of hair can form.

3 common presentation do pilonidal disease can occur:

1. Asymptomatic pilonidal sinus—a small pit or dimple is visible in the midline of the natal cleft.
2. Pilonidal abscess—acute inflammation and swelling develop as the pilonidal sinus becomes infected. This is usually away from the midline.
3. Chronically infected pilonidal sinus—if there has been an episode of abscess or infection then a discharging opening away from the midline in the buttock may be present.

Diagnosis

The diagnosis of pilonidal sinus is readily apparent due to the classical appearance. The differential diagnosis includes a congenital sacral pit or dimple, discharging dermoid sinus (which may be connected to the spinal tract) and very rarely tumours such as sacrococcygeal teratoma and rhabdomyosarcoma.

Look for the presence of “pits” in the natal cleft and lateral areas of swelling, erythema and discharge that represent acute or chronic abscesses.

Usually no further investigations are needed once the diagnosis is made clinically. In atypical cases an ultrasound and onward referral would be appropriate.

Primary Care Management

An asymptomatic pilonidal sinus with no associated infection can be managed conservatively. Give advice on regular cleaning of the area (a minimum of 2 baths/showers per week) and the prevention of hair accumulation. In hairy individuals, removal of natal cleft hair has been reported as being successful in preventing further disease in postoperative patients, but NICE guidance does not currently recommend hair removal in asymptomatic individuals.

If the sinus becomes infected and an abscess develops then urgent referral for incision and drainage of the abscess is required. An abscess should not be treated with initial antibiotics in primary care.

An initial course of antibiotics is warranted in a chronically infected discharging pilonidal sinus with cellulitis but no abscess. Co-amoxyclav or clarithromycin and metronidazole are appropriate first line treatments given the high proportion of anaerobes and mixed flora. Referral to a paediatric surgical center is then indicated for ongoing management.

When to Refer

- Pilonidal abscess should be referred urgently for incision and drainage under general anaesthetic
- Recurrent pilonidal abscess and chronically discharging pilonidal disease should also be referred to a paediatric surgeon with appropriate experience in the management of this difficult problem
- Atypical appearances and suspected pilonidal disease in prepubertal children should also be referred for further evaluation

After incision and drainage of a pilonidal abscess, consideration will be given to advice on hair removal and excision of the pilonidal disease. Likewise, chronically discharging disease will need further surgery.

From published studies the surgical management of pilonidal disease in children and adolescents has a higher recurrence rate than in adults and there is no clearly superior surgical strategy. Approaches rage from minimal excision of the affected pit through to wide local excision and primary or secondary wound closure. The choice of treatment will be influenced by the extent of disease and local expertise.

Chapter 27
Ingrown Toenails

Alison M. Campbell

Key Points
- Painful, relatively common condition impacting activities and quality of life
- Proper nail cutting, and foot hygiene are important preventative measures, but only have a small part to play once marked nail fold hypertrophy has developed.
- There is a high rate of recurrent problems even following surgical intervention

Introduction

In grown toenails are a painful, relatively common problem; affecting 3% of the population. Male patients predominate and 80% of cases are unilateral (affecting one aspect of one great toe nail).

Both congenital and acquired factors contribute. Rarely some neonates and infants have congenital deformities of the great toenails, these conditions are frequently self-correcting. Adolescents are more likely to require surgery. Although many may have a family history of toenail surgery; there is no clear genetic component. Indeed, nail shape has not been proven to be a risk factor. Congenital hallux deformity, hyperhidrosis, obesity and diabetes mellitus are risk factors for developing ingrown toe nails. However, tight footwear, poor foot hygiene, poor posture and gait and improper nail cutting are implicated in progression to ingrown toe nails. Repetitive or inadvertent trauma is also thought to provoke the process.

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In paediatric practice adolescents are more commonly affected than the younger age groups. A potential explanation is that adolescent feet perspire more, this results in softer nails. The nails split more easily, puncturing the nail fold, resulting in inflammation and infection.

Problems

Despite being considered a minor surgical condition, patients with ingrown toenails have lower quality of life scores on questionnaire than many with chronic health conditions. In addition, there is an impact on school work and participation in sport.

Recurrent problems, following both conservative and surgical intervention, are not infrequent.

Conservative Measures

Non-operative measures can be useful in the initial early stages of the disease process: education on nail care, soaking feet and antibiotic use when appropriate.

Neonatal ingrown toenail, where the nail margin has not overgrown the tip of the toe, tends to respond to conservative measures such as massage and not progress to inflammation and infection.

Infantile malalignment of the big toenail may also not progress and frequently responds to simple measures.

For prevention of problems on the contralateral side, education on nail care and foot hygiene is key.

Gutter splinting may be used by podiatrists to protect the nail fold from the ingrown toenail. However, by the time older children and adolescents present with ingrown toenails, surgical intervention is frequently required.

Surgical Management

Surgery for ingrown toenails in children may be provided by healthcare professionals other than paediatric surgeons. Surgical intervention tends to involve removal of a portion of the offending nail, although short-term follow-up studies do describe success with excision of the resulting hypertrophied nailfold (vandenbos procedure).

The most commonly performed procedure is wedge resection, usually with application of phenol to the nail bed. Procedures described range from simple nail avulsion to excision of the nail bed with surgical or chemical matricectomy. For these procedures recovery time, to return to full sporting activities, is usually 3 weeks. Pain is the main postoperative issue.

The re-operation rate for ingrown toenails is high (up to 1/3 of patients). In theory if matricectomy is incomplete, this could lead to new nail spicule along the lateral fold resulting in inflammatory reaction / recurrent IGTN.

Chemical ablation of nail bed (e.g. phenol) is associated with a decreased recurrence risk. However, it has been reported to increase risk of post-operative infection and prolong healing time.

Ablation of the nailbed with bipolar diathermy is an alternative technique for dealing with the nail matrix to prevent recurrence. However, over aggressive electrocautery may damage the underlying periosteum. This should be considered a possibility, if healing has not completed several weeks down the line post-surgery, and an x-ray of the distal phalanx performed.

There is no evidence that Manuka honey, antibiotics or various dressings speed postoperative recovery.

Indications for Referral to Paediatric Surgery

Failure of conservative management

Significant nailfold thickening and granulation suggests surgery will be required

Suggested Reading

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